THE INFANTILE CEREBRAL PALSIES

EIRENE COLLIS
Assistant to the Director of the Cerebral Palsy Unit, Queen Mary's Hospital for Children, Carshalton, and Honorary Adviser to the Cerebral Palsy Clinics of the Ospedale Maggiore, Crema, and of the Istituto Pio dei Rachitici, Milan, Italy

W. R. F. COLLIS
Director of Paediatrics, Rotunda Hospital; Physician, National Children's Hospital, Dublin; Chairman, National Association for Cerebral Palsy in Ireland

WILLIAM DUNHAM
Physician to the Department of Physical Medicine, Charing Cross Hospital; Consultant in Cerebral Palsy to the London County Council

L. T. HILLIARD
Consultant Psychiatrist and Physician Superintendent, Fountain Hospital for Mentally Defective Children, London

DAVID LAWSON
Physician Superintendent and Consultant Paediatrician, Director of Cerebral Palsy Unit, Queen Mary's Hospital for Children, Carshalton

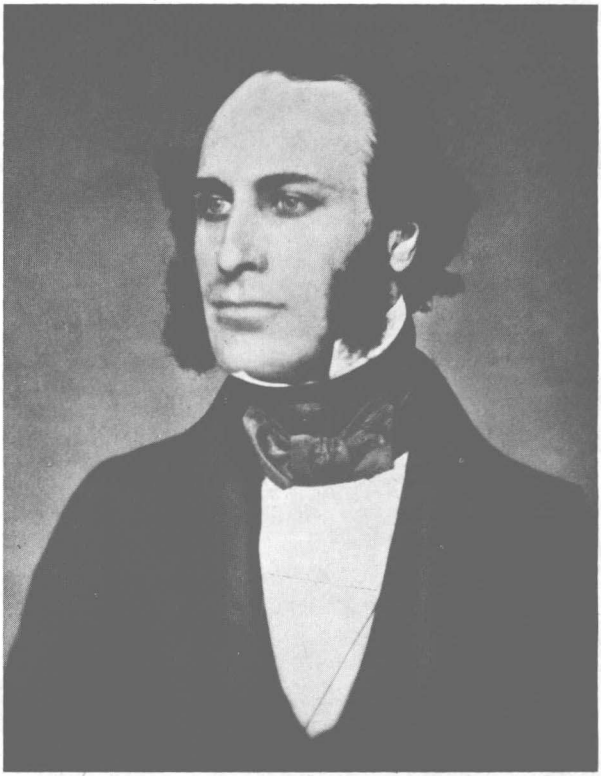
Foreword by

SIR FRANCIS WALSHE
Consulting Physician, University College Hospital and National Hospital for Nervous Diseases, Queen Square

1956
WILLIAM HEINEMANN · MEDICAL BOOKS · LTD
LONDON

First Published . . . 1956

Printed in Great Britain by Robert MacLehose and Co. Ltd.
The University Press, Glasgow



WILLIAM JOHN LITTLE (1810-1894)

LITTLE COMMITTEE FOR THE STUDY OF THE INFANTILE CEREBRAL PALSIES

THE Little Committee was formed one hundred years after the publication of Little's classical descriptions of cerebral palsy for the purpose of studying the clinical problems of diagnosis, prognosis, and management, and the administrative problems involved in training medical and other personnel for work in the field of cerebral palsy. The Committee enjoys the hospitality of Queen Mary's Hospital for Children, Carshalton, Surrey. The membership is as follows:

Mrs. Eirene Collis
Dr. W. R. F. Collis (*Chairman*)
Dr. William Dunham
Dr. L. T. Hilliard
Dr. David Lawson (*Secretary*)
Sir Francis Walshe

FOREWORD

THE wide and increasing interest on the part of the layman in this and other countries in the problem of what is now widely known as cerebral palsy in children has had, as could have been anticipated, consequences both good and bad.

Good, in that it has stimulated the renewed study of the forms of cerebral palsy, of its incidence in the community, and of its treatment on the part of the medical profession; and to have done this is at least to have taken a necessary further step towards that analysis of the disorders in question from which alone may we hope to achieve an optimum measure of benefit for the sufferers from these distressing afflictions.

On the other hand, less favourable consequences have flowed from this public interest, with its intense and quite natural desire for dramatic results: a desire untempered by an appreciation of the nature or complexity of the problems involved. Thus, ill-conceived projects are financed and precipitately launched without due and informed foresight, and a diversity of techniques of treatment, not invariably based upon sound physiological principles, not wisely used and thus not calculated to achieve the best possible results, are being increasingly employed. Only ultimate disillusionment and waste of human and material resources can come from all this, and one needs no prophetic gift to see that these consequences cannot be long delayed.

In the pages which follow, the essential, basic problem in the treatment of cerebral palsy is clearly expounded by the authors. It is a problem in applied neurophysiology, and we cannot emphasise too clearly that it requires from those who come forward to treat and to organise the treatment of the various disorders of movement involved, nothing less than a good measure of understanding of the neuromuscular control of voluntary movements. This control develops through known and recognisable stages during infancy and early childhood.

Since the child with cerebral palsy has a neural mechanism structurally defective, his acquisition of control of movement is subject to delay and to limitation which vary in character and severity accord-

ing to the extent and situation of the relevant lesion in the brain, for it must be remembered that the popular, but very imprecise, term 'cerebral palsy' covers a wide variety of disorders of movement and of posture, and does not refer to a unitary state in which the sufferers differ from one another merely in the degree of their disablement.

To study these defects and to analyse them in terms of the natural stages by which the normal infant acquires control of his movements and postures is a problem in child neurology. It calls for skilled diagnosis based upon trained observation directed specifically to this end, and, therefore, requires the long and patient study of the infant and child, normal and palsied. We cannot hope to detect the abnormal at its early stages unless we are familiar with the normal.

One has only to state the problem thus, for it to be realised how relatively small must be the field of competent and duly experienced observers: observers equal to the complexities of the problem and able to think physiologically. Need we wonder, therefore, at the amount of earnest but misdirected energy which we see around us, deployed in the therapeutic attack upon cerebral palsy?

Perhaps the criticism might be made that those who possess this essential knowledge have not adequately applied it in evolving rational modes of treatment. Whether this be a valid criticism or not, we may be sure that such modes of treatment will not be evolved without such knowledge, or by those who have it not.

William Little, the pioneer in the recognition and study of cerebral palsy, was fully aware of this and advocated the physiological approach to treatment as the only reasonable one. However, since his time, this, his really important lesson for us, has been largely forgotten, and quite other principles have come to dominate the climate of opinion as to treatment.

There has always been a dangerous temptation to think of treatment in terms of specific instruments or procedures which we 'try out' on this or that state of ill-health. A sage critic of this partial way of thinking has written that 'therapy is one of the routes by which medicine has been specialised, and by which the specialities have become vicious'. The specific treatment and what its proponents can hope to claim for it become more important than the patient. In few modern instances is this better exemplified than in the case of cerebral palsy.

Numerous therapeutic specialists now try out their own techniques on cerebral palsy, and too easily forget that their primary responsi-

bility and only rational aim is the training of the child for living, as it is expressed in the pages of this monograph. They forget the child as a person. They think of him as a spastic limb or as a deformity, as something to be fitted into the Procrustean bed of their method. If the dynamic training of the child from his earliest years, which is the theme so ably expounded in the following pages, is neglected, the child with cerebral palsy must sooner or later develop grossly abnormal patterns of movement and of attitude, and perhaps fixed deformities.

It is not surprising that the prevalence of these secondary consequences of a primary disorder of movement should have attracted the eager attention of the orthopaedic surgeon who, in his turn, tries out his own particular techniques upon them, without perhaps fully realising that he is not treating the primary and essential disorder of function, but merely the consequences of its neglect. In fact, surgical interventions and splintings are simply ineffective salvage activities that would never have appeared to be necessary had physiological principles of treatment been developed from the outset. I emphasise the distinction between salvage and treatment, since it defines a great deal of what passes for the treatment of cerebral palsy.

Of the various techniques of physiotherapy now so widely applied to cerebral palsy, we may again say that too frequently they are not based upon an understanding of the child's situation: but are procedures empirically tried out on him: a drill to one or other component of which it is hoped he may respond. In all this busy activity, the mother, the child's natural trainer, plays little or no part, is given no stimulus to co-operate, nor acquainted with the means by which she might fruitfully do so.

Doubtless, some parents, contemplating the position in terms of what they deem they have the right to obtain for their afflicted child, are willing to hand the child over to a team of white-coated technicians, and thus to place their own responsibilities in commission. In so doing they cherish a vain hope, for the mother is so plainly the natural and essential trainer of her child, whether it be normal or the subject of cerebral palsy, and no one can wholly replace her.

In short, the formal repertoires of orthopaedics and of physiotherapy, as we see them today, do not constitute a training of the child upon physiological principles, and are not optimally designed to meet his situation.

There remains yet another and a serious difficulty to be faced. This

is that the brain lesions that produce defects in the executive motor mechanisms of the brain, may also lead to mental defect in the child. Thus a child may have not only defective power of control of his movements from the one cause, but also a defective mental ability to learn to use what he has.

Thus the task of diagnosis in any case presented as one of cerebral palsy consists in determining whether one or both of these factors is present, and in assessing the relative importance of each in producing the total disability of the child to express himself in movements.

How complicated a problem of diagnosis is here involved needs no emphasis, or how fruitless treatment may be when not based upon accurate assessment and a realistic appreciation of what is possible.

Upon this aspect of the situation the public is woefully uninformed, as indeed are very many technicians who handle these children, and it is not surprising that the anxious mother of a mentally defective infant finding that her baby is not passing its milestones in due time, that he does not hold up his head, sit up, stand or walk, or use his hands, or articulate when he should, easily concludes that her child has cerebral palsy, and is resentful when the true nature of the situation is pointed out to her. Yet a considerable proportion of children referred to cerebral palsy clinics are, in fact, cases of primary amentia, or of children with both cerebral palsy and some degree of mental defect.

Enough has been said to indicate the intricate considerations involved, and the many pitfalls and disappointments that beset attempts to meet the situation.

In this monograph, the authors have provided a penetrating description of the various forms of cerebral palsy, and have laid down the physiological principles by which they are to be recognised, and by which they must be treated, and they have discussed clearly the complicating factor of mental defect. But more than mere description is offered of the distortions of movements and of attitudes these children present: each aspect of the abnormality is considered against the normal patterns appropriate to the age of the child, that is, in terms of normal function. The confusion between the symptoms of lesions and normal activities so commonly found in the literature of clinical and of experimental neurology finds no place here.

All the authors have pooled their considerable experience in this monograph in the making of a clear and coherent picture of cerebral palsy and of the necessary principles of its treatment; a treatment, or

rather a training, based upon the recognition of each child as a person who has, as far as is humanly possible, to be trained for independent living by means carefully adapted to each child. Throughout these pages there is evident the fruits of the many years of close study and experience of the normal and cerebral palsied child by Eirene Collis, who has acquired a deep insight into this problem: a fact widely recognised, especially on the Continent, by those interested in this subject. She has provided the creative element to be found in these pages: and she has been admirably supported by the general medical experience and special knowledge of her fellow-authors.

This monograph should prove a landmark in the literature of its subject. It is eminently practical. No one can read it without feeling how much more hopeful than had seemed possible is the task of all who may, in the light of its teachings, undertake the humane and satisfying rôle of training the child with cerebral palsy.

F. M. R. W.

LONDON
May 1956

CONTENTS

CHAPTER I

INTRODUCTION

THE recognition of motor disorders of cerebral origin arising during ante-natal development or around the time of birth, and thus affecting motor activity from the cradle to the grave, dates from Little's description in 1853[1] of the disorder which bears his name. As is so often the case, the key features of the group of disorders were very clearly described by the first observer. Little placed much emphasis on the frequent association of the motor disorder with widely varying degrees of mental defect, and also assured us that if intellect were unimpaired, considerable progress could be made by the homely means of constant education and encouragement in overcoming the defect. He also appreciated the important part played by birth trauma, and particularly by anoxia, in the genesis of the disorder.

Because of the importance of Little's work, and because his observations and deductions have stood the test of time and are still, a hundred years later, the foundation of our approach to the problems of cerebral palsy, we open this book with a biographical account, with excerpts from his work.

The dynamic approach which we here describe follows from the pointers which he gave a century ago, and which were for so long ignored.

When dynamic management was neglected in the early years, the majority of these children developed abnormal patterns of movement leading to fixed deformities of the limbs. With the progress of orthopaedic surgery, attention came to be directed to the physical correction of the worst of these secondary deformities, but this treatment was wrongly directed, as the deformity was merely the end product of a pathological situation which it did nothing to remedy. With the failure of this form of treatment and because the multiple disabilities from which these children suffered seemed to impinge upon a number of rapidly developing specialities such as orthopaedic surgery, physiotherapy, psychiatry, speech therapy and orthoptics, in addition to

neurology, there has arisen a tendency to pool resources and to try to diagnose and manage these children by a committee, none of whose members has the whole child in view, and, indeed, few of whom may have any special experience or understanding of growing children and their particular problems. As a direct result, the child's mother, the natural 'therapist', tends to be crowded out by a galaxy of professional talent.

Unproven techniques of increasing theoretical and practical complexity, often involving complicated equipment, tend to become de rigueur, and periodic attendances at a variety of Out-Patient Departments to displace the day-to-day guided management by the parents in the home. For, blinded by the complexity and zeal of all this professional endeavour, the parents tend to feel that their amateur efforts are superfluous, and allow these periodic and intensive assaults to take the place of that persistent daily help the value of which Little well recognised. This situation is tending to lead to an intensive provision of the wrong kind of service.

The infantile cerebral palsies are above all else conditions in which accurate diagnosis can be based only upon careful clinical examination by those who have long studied both normal and abnormal child development and behaviour, and in which the best results can be achieved only if the day-to-day care of the child is left where it belongs — in the hands of the mother, working under their guidance. None of the so-called 'techniques' required is so complex that it cannot be carried out satisfactorily by the average mother.

Our purpose, therefore, is to describe the natural history of the various types of cerebral palsy, and indicate the diagnostic criteria and techniques of management which have arisen out of the study of hundreds of cases in a pioneer cerebral palsy unit of which one of us (E.C.) has been in day-to-day control since its inception in 1943.

We do not pretend that the majority of these children can be rendered indistinguishable from the normal, nor do we imagine that we have solved all the problems of management, of education and of placement in society. We do, however, suggest that Little was on the right lines when he wrote:

> I have had many of these cases under observation from one to twenty years and may mention as an encouragement to other practitioners that treatment, based upon physiology and rational therapeutics, effects an amelioration surprising to those who have not watched such cases.

We shall emphasise the following, which seem to us to be cardinal to a useful approach to the problems of cerebral palsy:

1. There is no radical cure of the anatomical lesion itself. The processes by which effective motor function is achieved depend not upon structural recovery but upon the adaptation of function of those parts of the central nervous system not destroyed by the lesion.

2. The only way, therefore, which lies open for an approach to normal function is through careful and prolonged education of the child in the effective exploitation of his residual function.

3. Because this depends, as does any other learning activity, upon the active co-operation of the child, his ability to learn is of paramount importance. To such an extent is this true, that the prognosis depends far more upon intellectual capacity and persistence than upon the nature, extent and severity of the motor lesion itself.

4. In a high proportion of cases, the brain lesion affects adversely not only motor function, but also intelligence.

5. As motor achievement of one kind or another is used in all methods of assessing intelligence, there could hardly be a more difficult clinical problem than the assessment of the relative extents to which motor, mental, and perhaps sensory disability have affected a child's progress, in a group of disorders in which all are likely to be affected to variable extents.

6. In approaching an afflicted child, two questions outweigh all others in importance:

(*a*) Is there any evidence of specific motor defect? If not, then the delay in motor achievement must have its origin in intellectual disability. The apparent abnormalities of motor function in mentally defective children free of cerebral palsy are, on close study, *clearly defects of mental direction rather than of motor execution.*

(*b*) If there is evidence of specific motor disability, what are the child's intellectual resources? Here the final judgment must be a clinical one, and can be made only after a study which may sometimes involve weeks of day-to-day observation of the child and his responses. Accurate assessment is often difficult, but it is possible to divide the children into three groups: those whose learning ability is clearly within the normal range, those whose learning ability is so low that even without their motor disabilities they would not be educable, and a large middle

group of limited intelligence who to a variable extent will repay months or years of patient teaching.

Three gross, and common, errors can be avoided:

(*a*) Labelling as hopeless mental defectives, children whose major disability is a motor one and who are capable of learning to make great progress.

(*b*) The opposite error of assuming that no child with cerebral palsy can be safely diagnosed as intellectually ineducable.

(*c*) Making a diagnosis of cerebral palsy in children whose delay or abnormality of motor behaviour is the result of a mental defect alone.

These three errors between them, have probably given rise to almost as much avoidable distress as the failure to secure the proper management of cerebral palsy itself.

7. Some of the most distinctive signs of individual types of cerebral palsy may take many months to show themselves, because it is only as cerebral function gradually matures that the lesion can show its distorting effects. It is in the earliest months whilst the nervous system is at its most plastic, and learning is easiest, that the greatest progress can be made. Hence it is of paramount importance to the child to make the diagnosis during this plastic phase, before he has become morally inured to a life of physical incompetence and dependence, and before he has become further incapacitated by the secondary deformities to which prolonged neglect, inactivity and harmful treatment inevitably lead.

8. The treatment of the infantile cerebral palsies is more akin to the bringing-up of a normal child than to the formal treatment of a disease. No technique in which the mother and child are inactive can replace the hour-to-hour, day-to-day moulding of physical activity which the mother, under appropriate guidance, can best give. It is always difficult and sometimes impossible to reconcile the requirements of formal education and of physical education in a child who has reached school age before the management of his cerebral palsy has been effectively tackled; for if the pre-school years are lost, they can probably never be made up.

The following is an outline of the succeeding chapters. Chapter II is a short account of the life of William Little, with extracts from his classic descriptions of infantile cerebral palsy. Chapter III defines the clinical terms used in the book.

Chapter IV describes the nature of our approach to cerebral palsy, laying stress on some of the diagnostic difficulties which must be overcome before one is in a position to work out the best way of helping each individual child. We stress the overriding importance of mental capacity as an arbiter of the extent to which each child is able to respond to help given. And we state our view that the presence of cerebral palsy in a young child should not result in the mother being relieved of her responsibility for bringing him up, for the average mother, working under guidance, is the most effective 'therapist' imaginable.

Chapters V to IX describe the clinical entities referred to in Chapter III. These chapters each follow a common pattern:

1. Natural history of the disorder.
2. A brief case-history of a typical case.
3. Descriptions of the management of a child at various stages, laid out in the form in which advice was given to the mother.

Up to this point the book describes an approach, and techniques of diagnosis and management which have been learnt in the hard field of experience over many years. In Chapter X we suggest the principles which should guide those who are engaged in designing cerebral palsy services.

As a means of putting these principles into concrete form, the problems are considered in terms of a dense urban population of one million in England.

With the increasing interest in cerebral palsy which has occurred in recent years there appears to us to be developing a widespread misunderstanding of the nature and size of the problem. The wrong questions are being asked, and irrelevant answers sought.

Our purpose in this volume is not to lay out ready-made administrative and clinical solutions capable of immediate world-wide application, but rather one of defining the problems so that they may be clearly viewed.

This is done mainly in terms of the experience of one of us (E.C.) in the pioneer unit at Queen Mary's Hospital for Children over the past fourteen years[3 5 6 7]. In this experience the other authors have to a varying degree participated.

The structure of this service has evolved slowly in the light of experience, and in a particular environment, but it has always been allowed to change along the lines which experience suggested.

We are confident that the principles learnt in this unit, and expounded in this book, are those which must underlie the management of cerebral palsy in whatever differing environments it may be undertaken.