CEREBRAL PALSY OF CHILDHOOD IN EDINBURGH: AN ASSESSMENT OF ITS INCIDENCE AND A DESCRIPTION OF THE CLINICAL FINDINGS IN 208 PATIENTS.

by

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CEREBRAL PALSY OF CHILDHOOD IN EDINBURGH: AN ASSESSMENT OF ITS INCIDENCE AND A DESCRIPTION OF THE CLINICAL FINDINGS IN 208 PATIENTS.

This essay describes some of the results obtained from research carried out during the tenure of the George Guthrie Research Fellowship in the Department of Child Life and Health of the University of Edinburgh in 1952 and 1953. The purpose of the research was to ascertain the incidence of cerebral palsy amongst children in Edinburgh and to define some of the factors which were important in the aetiology of the condition.

What follows is a description of the methods of the survey, and the classification of cases that was adopted. The clinical findings of 208 patients who were resident in Edinburgh and suffered from cerebral palsy are presented. The impairment of normal activities which results from cerebral palsy is emphasised. The incidence of cerebral palsy in Edinburgh is discussed and some attempt made to assess the burden which cerebral palsy causes to the community. Finally the facilities available for the medical treatment, education and provision of work and the facilities which are desirable are considered. In this essay the aetiology of cerebral palsy is not discussed.

The essay does not pretend to be a comprehensive account of the clinical findings encountered in children with cerebral palsy. Rather, facets of the clinical

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picture which have interested the author and appear to be of importance to him are considered. Thus what may be considered a disproportionate amount of space is devoted to the clinical findings at different stages of diplegia. The justification for the attention devoted to them lies in the present delay in the diagnosis of diplegia, which is on average almost two years after the first manifestations of the conditions have occurred. Moreover the clinical findings in the early stages of diplegia have not been considered very fully in the literature and they interested the observer greatly.

Nevertheless it can be claimed that the clinical abnormalities which result in the most severe functional impairment of voluntary control of the limbs have been considered fairly fully. Some attempt has been made to relate the various disabilities suffered by patients with cerebral palsy to their capacity to receive education and later to work.

The Scope of the Survey.

The survey was confined to children resident within the boundaries of the City of Edinburgh in order that an accurate figure for the total child population might be available and because the child population within those limits was of a convenient size for study. Initially it was confined to children born between 1943 and 1952, children of school age, because these were more accessible for examination than preschool children or those who had left school. Later, however, it was found possible to extend the survey to include children below school age and though the study of these children was less comprehensive than of those at school, information of great interest was obtained. A small number of children over the age of 15 who had left school were also examined in order that some insight might be gained into the problems of employing children with cerebral palsy, but they are excluded from the series of cases to be presented.

The survey included all children resident in the City of Edinburgh born between the years 1938 and 1952 inclusive, whether they were in Corporation schools, private schools or were considered ineducable, and whether at home or in institutions.

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An effort was made to examine all children in whom there was the least suspicion of cerebral palsy being present. Very large number of children were examined in schools, at home, in institutions, hospitals and clinics. In every child confirmed to be suffering from cerebral palsy on examination, very full clinical details of the history and examination were recorded. Efforts were made to obtain details of the child's birth and progress from doctors and hospitals and of educational progress from schools.

As a result of the investigations it was possible to make an assessment of the incidence of cerebral palsy in childhood in Edinburgh. At the same time a picture was obtained of the handicaps shown by children suffering from the condition and the means available for treating them.

Methods of the Survey.

Various methods of estimating the incidence of cerebral palsy in childhood were considered. Obviously a population of more than 100,000 children was too large for the individual examination of every child to be possible. Sampling survey was impracticable because of the relatively low incidence of cerebral palsy in the community. Assuming that there are /

are two cases of cerebral palsy for every 1,000 children of school age in the community, as suggested by the British Council for the Welfare of Spastics, a sample of at least 15,000 children would have had to be examined for reliable incidence figures to be obtained. This would have entailed arranging for the examination of approximately one sixth of the population of school age in Edinburgh. This was an impossibly large and complicated task in the circumstances of the investigation. Even if such a sampling survey had been accomplished it would have resulted in approximately thirty cases of cerebral palsy being discovered. This number was clearly inadequate for it to be possible to consider aetiological factors which they showed as being representative of all types of cerebral palsy.

It was decided to attempt an estimation of the incidence of cerebral palsy directly by ascertaining as many cases as possible from all sources and all ages to 15 inclusive. It was realised that, by using this method of investigation patients suffering from cerebral palsy could very easily be missed. On the other hand because of the highly organised Maternity and Child Welfare Services, school medical and hospital services and the presence of a school for children with cerebral palsy, Edinburgh was a particularly suitable place for such /

such an attempt to be made. Owing largely to propaganda by the Scottish Council for the Care of Spastics there was great awareness of the problems of cerebral palsy amongst parents, teachers, health visitors and medical men. Therefore cases tended to be reported shortly after the child reached school age, if not before. Moreover with a population of approximately 100,000 children born between 1938 and 1952 and resident in Edinburgh, a total of about 200 patients with cerebral palsy might be expected. The study of this number of cases would give enough information for conclusions about aetiology to be reasonably representative, yet the numbers of patients was not too large to make full individual examination impossible.

Obviously the success of an attempt at direct ascertainment of cases of cerebral palsy would depend on the co-operation of school, hospital and other medical authorities, especially in giving access to records and children in their care, and on the permission of parents to the examination of their children being obtained. In these particulars the investigator was most fortunate.

Means of Ascertainment.

In the initial stages of the survey three lines of /

of investigation were pursued in order to simplify later and more accurate ascertainment of cases.

Firstly very large numbers of medical records from different sources were inspected and lists made of children in whom the existence of cerebral palsy seemed even a remote possibility on the evidence available.

Secondly children suffering from conditions known to be associated with cerebral palsy, such as epilepsy, mental deficiency with clumsiness, marked overactivity, were listed. At the same time children who had suffered from conditions which could lead to cerebral palsy were noted. These included birth injury, rhesus incompatability, encephalitis, head injury and meningitis.

Thirdly, arrangements were made for screening inspections and brief examinations to be made in groups of children in whom there was likely to be a high incidence of cerebral palsy. Children were seen, for example, in institutions for the mentally defective, schools for the mentally and physically handicapped and in prematurely born infant clinics.

These three means of approach will be considered in turn in more detail.

1. Listing of possible cases of cerebral palsy.

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The following sources provided the facilities for the listing of cases diagnosed as suffering from cerebral palsy or thought possibly to be suffering from the condition by the investigator.

Corporation Schools. Access was granted to the medical records of the Corporation School Medical Service. These were invaluable because they provided details of very full examinations of those groups of children in which a high proportion of cerebral palsy might be expected to occur, the schools for the mentally and physically handicapped and children classified as ineducable. The details available of children in normal schools were less full but quite adequate in nearly all cases to make it clear if a suspicion of the existence of cerebral palsy should be entertained or not. In all, approximately 6,000 case notes were inspected in various schools. Of more value than the inspection of the records of the School Medical Service was the co-operation of the Medical Officers, nurses and teachers. Many cases thought to suffer or possibly to suffer from cerebral palsy were reported by them and though the majority of these cases had been listed following scrutiny of the medical notes, their reports served as a very valuable check. In the normal schools a large number of cases were reported by School Medical Officers, though usually also ascertained by other /

other methods.

Private Schools. Approximately 5000 children in Edinburgh attend private schools and since a number of these were likely also to be private patients and thus avoid ascertainment through hospitals and health visitors, it was felt necessary to follow up as many as possible.

All Head Masters and Mistresses of private schools and nursery schools in Edinburgh were contacted and asked for any information they could give about possible cases of cerebral palsy amongst their pupils. Of 55 schools that were contacted, all but two gave the information requested.

Maternity and Child Welfare Service. Free access to case notes, clinics and Medical Officers of the Service was most readily given. All health visitors and matrons of nurseries were contacted and gave details of children whom they suspected of suffering from cerebral palsy. The Medical Officers of the Department referred a number of children for examination from their routine clinics.

The co-operation of the Service was especially valuable because of the relatively fewer sources of information available about possible cases under school age.

Public Health Department. /

Public Health Department. Access to the records and patients attending clinics of the department was freely given and in addition the officers of the department's clinics were most helpful in reporting cases and possible cases of cerebral palsy. Cases reported from the orthopaedic clinics were especially valuable as they tended to be older children with fixed deformities who had ceased to attend other clinics.

Hospitals. All hospitals in Edinburgh concerned with the diagnosis and treatment of children's disorders, of whatever type were contacted and in every case access to notes and patients was readily granted by those in charge. Staff was questioned and notes reviewed to find possible cases in infant clinics, orthopaedic, neurological, neurosurgical, general medical, general surgical and infectious disease wards and clinics.

As many staff were contacted as possible and the heads of departments were most co-operative, in reporting cases, arranging for cases to be seen at convenient times and introducing the investigator to the parents of affected children.

In all hospitals the physiotherapy and occupational departments were contacted and gave information about possible cases.

Scottish Council for the Care of Spastics. /

Scottish Council for the Care of Spastics. This organisation gave free access to its school and in and out patient records, which contained much information about a large number of cases of cerebral palsy. Quite apart from what was learnt from case notes and the examination of pupils in the school, much information about cases was obtained from the staff, who were always most helpful.

General Practitioners. It was originally intended to approach all Edinburgh general practitioners and request information about possible cases of cerebral palsy in their practices. As a first step personal letters were sent to twenty-five doctors known, at that stage of the investigation, to have at least twenty-eight cases of cerebral palsy in their practices. Replies were received from nineteen of the doctors. They gave the names of two cases, both already known from other sources. It was therefore decided not to write, in future, specifically to ask for the names of cases of cerebral palsy in their practices, but whenever a doctor was asked for permission for one of his patients to be examined, a request for information about any other cases knownto him was enclosed.

Physicians and Surgeons. The response from physicians and surgeons dealing with cases of cerebral palsy to the request /

request for information about possible cases was very good. Cases were notified and times of attendance at their clinics given very frequently by extremely busy men. A number of consultants gave introductions to their private patients, whom it would have been very difficult to contact in any other way.

Other sources. A number of cases were reported by investigators engaged on other surveys who incidentally encountered cases of cerebral palsy. Cases were reported as a result of an investigation of cases suffering from mental defect (Dr. H. Provis), a survey of premature infants from the point of view of retrolental fibroplasia (Dr. J.D. Kerr), a survey of premature births from the point of view of social conditions (Dr. C.M. Drillien), as a result of an investigation of growth in childhood (Dr. John Thomson), as a result of a survey of the after effects of meningitis (Dr. J.W. Farquhar, and of an investigation into cases of birth injury (Dr. Charles Balf). A few cases were reported from speech therapists, psychologists and nurses attached to clinics, hospitals or schools and child guidance clinics.

2. Ascertainment of Children with related disorders.

Since mental deficiency, epilepsy, overactivity, arthrogryposis, talipes equino varus were found relatively /

relatively frequently to occur in patients with cerebral palsy, lists of patients with these conditions were made and their notes inspected. Frequently inspection of the notes was sufficient to rule out the possibility of cerebral palsy being present, but many others were examined. For example a large number of those with mental defect had been examined with great care in a special clinic and where it was stated, as a result of a relatively recent examination that neurological findings were normal, this opinion was usually accepted. On the other hand a large number of mental defectives were stated to be clumsy or "slightly ataxic" by other clinicians and all these cases were examined. Similarly though the majority of the 300 cases of epilepsy were examined, if only briefly, a small number where examination would have been difficult and in whom neurological examination was stated on good authority to be entirely normal, were not seen.

In addition to listing children with possible co-incidental abnormalities, children who had suffered from conditions known to lead to cerebral palsy in a proportion of cases were also noted. These included, birth injury, rhesus incompatability, prematurity, encephalitis, meningitis, cerebral thrombophlebitis, head injury, severe /

severe dehydration and cerebral complications of infectious diseases. It was not possible to investigate these categories as fully as was wished.

Prematurity. Children born prematurely between the years 1948 and 1952 inclusive had been thoroughly followed up in a survey concerned with ascertaining the incidence of retrolental fibroplasia (Dr. J.D. Kerr) and any child suspected of suffering from cerebral palsy had been referred. Since children born prematurely before 1948 were of school age it was felt not to be necessary or desirable to follow them up as a special category.

Birth Injury. Children who had suffered from birth injury in Edinburgh hospitals had been examined in 1949 by Dr. Charles Balf and the results of his investigations were available. Since children resident in Edinburgh who had suffered from birth injury or shown abnormality in the neonatal period almost routinely attended infant clinics, it is unlikely that many cases showing persistent neurological abnormality, born since the previous survey, have been missed. All known cases in whom persistent neurological abnormality had been noted by clinics or health visitors were examined.

Rhesus Incompatability. It was felt undesirable to investigate the effects of rhesus incompatability fully because /

because it would have been impossible to correlate the degree of nervous damage sustained to the haematological findings and the clinical findings at the time of parturition, owing to shortage of time. Since cases of rhesus incompatability routinely attend infant clinics following their discharge from hospital, and since the notes of all cases showing rhesus incompatability were inspected and all children noted to show persistent neurological abnormality examined, it seems likely that relatively few cases of cerebral palsy due to rhesus incompatability were missed. In a small scale survey of children who had suffered from the effects of rhesus incompatability in the Elsie Inglis Hospital, only one slightly affected child not known from other sources, was discovered out of 35 children examined.

Encephalitis. All the case notes of children noted as having suffered from encephalitis, whether as the result of infectious disease or other cause, were inspected and in any where there was the least suspicion of residual paralysis, the children were examined.

Meningitis. At the time of this investigation a survey was being made by Dr. J.W. Farquhar and Dr. I.C. Lewis into the after effects of septic meningitis in childhood. Unfortunately they were unable to complete their survey, but the list of patients they obtained was utilised and all /

all were examined unless there was very clear evidence that neurological findings were normal. Of the few patients seen during the meningitis survey, two showed cerebral palsy. Both these cases had been noted to be cases of cerebral palsy as a result of information obtained from other sources.

Cerebral throbophlebitis and abscess. All cases noted to have suffered from these conditions were examined unless recent full examination had shown normal neurological findings and arranging for the child to be seen was difficult.

Head Injury. Cases in which residual neurological abnormalities following head injury had been noted, were examined. Cases known to have suffered head injury in which no note had been made of residual abnormalities were examined unless stated by to show no abnormality by the staff of the neurosurgical unit. Since the majority of cases of head injury were seen by them relatively few cases who had suffered from head injury without evident sequelae were examined during the survey.

Cerebral complications of other diseases. Cerebral complications following non specific fevers, exanthems, dehydration, congenital heart abnormalities, and diabetes were noted and the patients examined if any persistent disability /

disability seemed likely to be present.

3. Active ascertainment in groups of children where a high incidence of cerebral palsy might be expected.

In contrast to the two previous means of ascertainment described where information at second had was utilised, large numbers of children in groups likely to show a high incidence of cerebral palsy were seen personally. After a few months during which experience was gained, it was found most convenient and rewarding to watch children at play in order to select those in whom further examination was desirable. This apparently crude method of ascertainment was more effective than might have been expected. In one school where it was possible to examine 60 children and in an institution where over 100 children were examined no additional cases to the nine diagnosed on inspection as suffering from cerebral palsy were found on later full neurological examination. The usual procedure adopted was to inspect the available medical notes and ask teachers and staff for details of any children suffering from marked clumsiness, inco-ordination, fits, marked writing difficulty, left handedness or other symptoms suggesting the presence of cerebral palsy. The children were then inspected at play for as long as possible at a time or on several occasions and then in the classroom, where dyskinesis was frequently more readily noted. All children showing abnormalities /

abnormalities suggesting that they might be suffering from cerebral palsy were noted and arrangements made for their thorough examination.

Inspections of this type were carried out in all the Corporation Special Schools for the mentally and physically handicapped, a number of private schools for the physically and mentally handicapped and blind, institutions for the mentally defective and in one nursery school.

Inspections were also carried out in a number of infant welfare clinics on a more limited scale.

Results of the preliminary ascertainment.

As a result of the preliminary surveys described above it was possible to list the cases it was thought desirable to examine further. Cases were divided into probable cases and possible cases of cerebral palsy. The former were those diagnosed by other clinicians as suffering from cerebral palsy and those thought to be cases of cerebral palsy by the observer as a result of his inspections. Possible cases were those who had suffered from any condition likely to lead to cerebral palsy, or showing symptoms or conditions known to be associated with cerebral palsy unless this could be excluded as a result of previous reliable examinations.

As a result of the preliminary ascertainment, 484 children were classified as possible cases of cerebral palsy /

palsy and 165 were classified as probable cases of cerebral palsy.

Further ascertainment and diagnosis of children suspected of having cerebral palsy.

All cases classified as probable were examined with a few exceptions who refused full examination. In these it was necessary to utilise the notes of examination made by other observers.

It was impossible or impracticable to examine all cases classified as possibly suffering from cerebral palsy, for a variety of reasons. Between 80 and 90 patients, in fact, were not examined. In thirty of these, chiefly cases of rhesus incompatability or mild epilepsy, it was found impossible or undesirable to contact the patient directly or indirectly to ascertain the presence or absence of cerebral palsy. In the remaining cases, contact, direct or indirect, was made with the patient and it was possible to be fairly certain that cerebral palsy was not present.

Thus of the 484 patients classified as possible cases of cerebral palsy, approximately 400 were examined. In most of these the examination was brief and concerned solely with confirming that palsy was not present. In about 80 or 90 patients personal examination was not made, and in thirty of these no direct or indirect contact was made at the time of the survey. In the remaining /

remaining 50-60 patients second hand opinions as to whether or not cerebral palsy was present had to be accepted.

Thanks to the co-operation of school medical officers it was possible to examine most of those whom it was desired to see, at the time of school medical examinations, frequently arranged especially for the benefit of the investigation. In all, approximately 420 children were examined in the schools. In the examinations of the possible group no notes were taken of those found not to suffer from cerebral palsy, their names were merely erased from the list of possibles. Full notes were taken of all those in whom cerebral palsy was thought to be present or in whom it was suspected even though these children were all seen on subsequent occasions.

Other cases were examined in institutions for the mentally defective, private schools of various types, hospitals and clinics, corporation nurseries and departments of occupation physiotherapy. Not only did this technique of attempting to arrange the first examination of the child away from home save the parents, (and the investigator), much time, but it also enabled contact to be maintained with a large number of clinicians and therapists and their interest in the investigation sustained. It was an added advantage that the children were seen in surroundings different from those of their own homes on at least one occasion.

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A number of patients, especially those under school age, those classified as ineducable and those having home teaching, were examined only in their homes.

Following the first examination arrangements were made with the parents to re-examine and take the histories of, those in whom cerebral palsy had been diagnosed. Most frequently these children were visited at home but a proportion were examined at the time of their routine attendance at various clinics, in the Department of Child Life and Health and sometimes in hospital. Even when, for convenience of the parents, the child was examined away from home an effort was always made to visit the home on at least one occasion. It was possible there to observe the family's reaction to the handicapped child, the behaviour of the child at home and frequently other relatives than the mother were contacted. On three occasions observation of relatives led directly to the recognition of familial neurological disorder, which would not otherwise have been evident.

Methods of Examination.

The methods used in history taking and examination will be described briefly.

History. The history was taken, on all possible occasions from the mother because of her much greater knowledge of the child's early, and especially neonatal history.

Whenever /

Whenever possible the mother was questioned on at least two occasions, preferably in different situations. The order of history taking, though of necessity somewhat varied according to circumstances was generally as follows.

A brief family history was taken first. During this information was elicited about the number of people in the family, the patient's place in the family and how he compared in achievement and milestones to his siblings.

The patient's first abnormality was noted and the mother's opinion as to the cause of this. This led naturally to questions about pregnancy and delivery and the neonatal course. At this stage questioning became more detailed and a very full history was taken of the child's milestones, growth, handedness, dexterity, intellectual development and ways in which his early development and behaviour differed from the normal. Such detailed questioning at this stage allowed one to slow the tempo of the interview and facilited the mother's thought and the investigator's notetaking. Following the history of the child's early development, the history of the appearance of the first abnormality was taken in more detail and the course of the disorder was followed in detail. This part of the history was usually /

usually taken more by encouraging the mother to talk than by close questioning; when her flow of talk had stopped then detailed interrogation was resumed using her story as a basis. In practise this involved a considerable amount of direct questioning in order to elicit points which would not seem of relevance to the mother. A frequent example was, "When he came back from hospital did you find any difficulty in washing him which you had not noted before?" "No, of course not, he was just the same". Direct question. "Did you do anything to the right hand before you washed it". "Yes, I had to open it, he always held the fingers closed, though not the other hand". Following such detailed questioning about the origin and development of the child's disorder, questions were asked about his behaviour, illnesses, general educational progress and sociability. Details were then obtained about the history of the family with the emphasis on the mother's obstetrical history and the occurrence of other mental or nervous disorder. Depending on the degree of the mother's co-operation, direct questions might be asked about the occurrence of mental defect and aberration in relatives or this left until further interviews. Direct questions were asked about consanguinity of the parents. Finally a history of the child's treatment was obtained, with the names of doctors, therapists, clinics, schools, teachers and others who had come into contact with the child and permission /

permission obtained to utilise any records obtained.

Following this interview with the mother another appointment was made to see her, either at home, at one of her routine clinic visits or by special arrangement. After the mother had been seen, as many relatives of the child as possible were seen, especially to obtain an accurate history of the mother's relatives.

In all cases attempts were made to obtain details of the mother's pregnancy with the patient, his birth and neonatal period, from additional sources to the history elicited from the mother or relatives. In 98 cases it was possible to abstract full details from notes of maternity hospitals. Since they relied to a greater extent on memory, details obtained from general practitioners, nurses and nursing homes were relatively scanty and less reliable. Occasionally doctors were able to supply very full details of deliveries which had occurred many years before, by reference to their notes.

Notes of infant welfare clinics, hospital clinics, wards, general practitioners, therapists and specialists who had examined the children were requested in all cases. The information was helpful in checking the accuracy of the history obtained from mothers and relatives, in obtaining a picture of the development of the child's disorder as well as giving an account of the actual causal condition in many instances.

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In all possible cases details of the child's progress at school were obtained, particular emphasis being placed on ascertaining if specific learning difficulties, reversing letters or words, aphasia, or speech defect had been encountered. The child's reaction to his fellows and his ability to play with them was noted and in many cases details about the educational progress of siblings was obtained. Frequently psychological testing of children with cerebral palsy had been very full and was made available. The co-operation and interest shown by teachers in both corporation and private schools was great.

The Examination.

The examination of the child was carried out on more than one occasion in different circumstances in as many instances as possible. The advantages of this were that it was possible to assess the influence of changes in environment on the child's behaviour, and to gain the child's co-operation, if not at the time of the first examination, in almost all cases by the time of the second or third. Especially in dyskinetic patients changes in findings were discovered. Abnormalities missed on initial examinations could be recorded.

The methods and detailed order of examination varied greatly in different cases and circumstances. The child was /

was weighed and measured for height, difference in limb lengths, trunk symmetry, and head circumference. Any skeletal abnormalities and obvious congenital defects were then noted. The child's demeanour, general behaviour co-operation, apparent intelligence, ability to answer questions and respond to commands were commented on briefly. Note was taken of any speech abnormality that was present and his ability in social activities, reading and writing. A brief general physical examination was made routinely and any abnormalities recorded. A detailed neurological examination followed. It included tests of visual acuity and fields, but not detailed testing of hearing, taste or, in young children palatal and tongue movements. The detail of the examination varied greatly with the age of the child and in practise sensation could be tested fully only in children with mental ages of 7 years or more.

Greatest attention was paid to examination of the motor system, the child's postural reflexes, tendon jerks and muscle tone being noted in great detail because they all could be tested in even the youngest.

Some attempt was made, in al cases, to assess the degree of functional impairment suffered by the child and his ability to sit, stand, walk with and without support, steadiness and manipulative ability were tested. In all instances the child's ability to feed hi self, dress /

dress himself, handle small objects, manipulate large and small objects, write and co-ordinate his movements in practical situations was noted. From this a far better idea of his capabilities was obtained than by giving standard tests like picking up pins or marbles or completing formboards.

Special examinations. Intelligence quotients were not obtained routinely for the purposes of the survey because most of the children of school age had already been tested and estimates of their intelligence were available. A number of children, especially those under the age of 5 were tested specially by psychologists attached to various schools, departments and hospitals. In a number of cases testing had never been performed and was not requested, either because the child was obviously ineducable or because his abilities were very apparently of a very high order.

Routine electroencephalograms were not performed because of the time necessarily lost to children of school age in visiting the Department of Surgical Neurology, where they were performed. In a large number of children they were available however, and in others, where it was felt they would be of value, they were obtained.

Rhesus blood grouping was not routinely performed, but /

but in all cases where any doubt about rhesus incompatability was felt they were performed on request by the S.E. Regional Blood Transfusion Service.

Radiographs were requested in very few cases. This was partly due to the fact that a special journey would have had to be made by the children to have them made. It is much regretted, in retrospect, however, that more radiographs were not obtained especially from the point of view of estimating bone age and of obtaining permanent records of skull abnormalities.

Fortunately from the point of view of the survey, a number of children had been admitted to hospital and air encephalographs obtained. These and other hospital examinations were readily made available to the investigation.

Some difficulties encountered during the survey.

The difficulties encountered during the survey were somewhat unexpected and some attempt to note their nature and occurrence was made as the investigation progressed.

Ascertainment. Difficulties in ascertainment will be discussed in detail when the measure of accuracy attained in the assessment of the incidence of cerebral palsy is considered. The difficulties were less than had been expected /

expected owing to the very great measure of co-operation given by clinicians, clinics, schools and hospitals. Since the ascertainment was very largely dependent on information received from others, however, failures resulting from human fallibility were encountered. For example, though clinicians frequently intended to notify cases they had seen to the investigator, they sometimes forgot, or noted the name on a piece of paper and lost it. As close contact as possible was maintained with all such clinicians and to some extent the likelihood of cases being missed was lessened by the large number of sources from which cases were notified.

Considerable time and trouble was also expended on obtaining the correct addresses of children who had moved since the time of the making their medical records, in some cases dated 10 or 12 years earlier. Some times records were inadequate and it was necessary to place a child on the possible cerebral palsy list because of inadequate information about him, rather than any positive finding. This occurred most frequently with children suffering from epilepsy or mental deficiency, and quite as frequently in those who had been seen in hospital or elsewhere.

Very free access to children in schools of all types, institutions, hospitals, and the homes of those who could not be seen elsewhere, so that surprisingly little difficulty was encountered in the inspections of large groups of children.

History taking. /

History taking.

It was found a great advantage to question more than one member of the family, especially about family history. On one occasion the father remembered an attempt at abortion, "forgotten", and actually denied later by the wife.

Whenever possible the mother or guardian of the child was questioned on at least two occasions, preferably in different situations. It was rapidly realised that practically every parent has some feeling of guilt, expressed or suppressed, about her affected child. Consciously or subconsciously such guilt feelings were bound to colour the mother's account of her child's disorder. Sometimes this led to the deliberate suppression of relevant personal history, but more frequently to more subtle, unintentional, subconscious forgetting of details. At a third interview one mother remembered falling from a step ladder on which she had been working when supposed to be taking things easily at the 38th. week of pregnancy. Memories of pregnancy and delivery seemed to be especially liable to be coloured by this form of subconscious suppression of information. Thus one intelligent ex nurse with a passion for psychological literature insisted that her pregnancy had been completely normal, in spite of hospital records of an admission for hyperemesis. /

hyperemesis. She added, "He was a wanted child, - now why did I say that."

Though careful questioning obviously went some way to revealing such half forgotten, half suppressed facts, a much more potent means of obtaining them was by letting the mother feel at her ease and talk. Almost inevitably as a result the emotional tension caused by her suppression of facts and feelings of guilt, relevant facts emerged eventually. Frequently there was great relief when they did. In contrast some mothers could not be induced to talk and others would have required too much time, even with a generous allowance, to produce any fact of significance.

Deliberate suppression of facts was common. One rapidly developed a sense of "skeleton in the cupboard", however, and in many cases a second or third interview arranged on different pretexts resulted in a change of attitude. No attempted abortions were admitted spontaneously before the interview time had exceeded one and a half hours. Only one was admitted under two hours and three under two and a half hours. It was similarly found that illegitimacy, especially in women subsequently married and "respectable", was concealed for many hours.

Deliberate lying was encountered several times.

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In two cases stories had been concocted with the obvious idea of giving enough information to justify divorce proceedings. In one of these a polite request for the truth after one hour's rigmarole produced tears and a much more credible story. Probably the most subtle alteration of fact was the subtracting deliberately of 12 months from the age of a slightly backwards boy who was "Doing much better at school since we've come to Edinburgh, thank you."

Apart from deliberate and involuntary suppression or distortion of facts many women were genuinely ignorant of details concerning their children. In many cases this was due to ignorance of even the simplest facts of normal child development, especially liable to occur since so many of the children were first born. Ignorance of what constituted normal pregnancy and delivery was especially prevalent. Two women accepted a spot of bleeding now and then in the later months of pregnancy as normal. Where ever possible recourse to hospital notes was made, though occasionally, as in the case of one child born in a hospital lavatory, the mother's story was so circumstantial and confirmed from other sources, that it was accepted, in face of conflicting evidence from the hospital.

Ignorance, failure to notice abnormalities and poor memory /

memory were much commoner in mentally defective parents, frequently of mental defective children. The histories obtained of such children were often woefully incomplete.

Whereas the history of the first born was often rendered difficult to interpret because of the mother's ignorance of what was normal and what was not in her child, the mothers of very large families had ceased to take such a close interest in their children's early months and their failure to observe was frequently marked.

Other reasons for defective histories were the child's separation from the mother and guardians, either permanently or temporarily. Frequently adoptive or foster mothers knew nothing of the early days of their children and the details of some in institutions were very scanty. Though in theory separations due to hospitalisation were compensated for by the availability of hospital notes, in fact these were found of much less value than a continuous story from the mother or guardian of the child.

It was found impossible to trace 5 mothers and the histories were inadequate and derived from other notes. Co-operation was refused directly, not merely by default, in 2 cases. The histories regarded as adequate in 165 cases, somewhat inadequate in 31, and grossly inadequate in 12. Of the histories regarded as inadequate, in 28 cases the parents were known to be mentally defective or backward.

Examination.

Access to children was not obtained in two cases because of parental refusal to co-operate. It was possible to ascertain that one of these children was a non-identical twin girl aged 5 with spastic paraplegia, strabismus and mental retardation by a brief informal examination as the mother dilated on the subject of the wickedness of doctors. The other child was a severe right hemiplegia in a boy aged 15 and notes of his birth were traced but no other details obtained.

In general it was possible to obtain access to the patients with cerebral palsy and most parents welcomed examination because they felt it showed some interest was being taken in the patient and that some treatment might be possible, though it was always emphasised that the investigation was not concerned with treatment.

Difficulties of examination were due to poor co-operation on the part of the child or to the nature of the condition from which the child suffered.

Poor co-operation was due, most commonly, to the child being very young, mentally defective, afraid of doctors, or behaving badly due to faulty handling or organic behaviour disorder. In general it was possible with patience to achieve co-operation sooner or later so that examination could be adequately performed.

Difficulties /

Difficulties due to the disorder from which the child suffered were more difficult to overcome. Mentally defective children, for example, were frequently very clumsy and appeared to be ataxic. To what degree the clumsiness was due to organic neorological disturbance, in distinction to generally poor development of motor skills, a manifestation of the mental deficiency, it might have been possible to determine by full tests. Since the child was incapable of these, great difficulty in distinguishing these border line casds was encountered. A number of dyskinetic patients showed their involuntary movements only under circumstances of great stress and occasionally pictures of gross associated movements described in detail by others, were not found on examination.

Patients having epileptic attacks also presented considerable difficulty as the findings following attacks were sometimes much changed, occasionally for a week or longer. It was often necessary to examine these patients on 3 or 4 occasions before a reasonably firm opinion could be reached about the degree and nature of their disabilities.

It was particularly difficult in mentally defective patients with long standing disease, accompanied by severe contractures, to obtain any clear idea of what lesions had originally been present and necessitated the recumbency which had resulted in contractures. These patients /

patients were an exception to what was generally found; that the longer time spent on examining the patient the clearer became the picture of his disorder.

The greatest difficulty was encountered in the examination of sensory findings in the very young and the very defective. Frequently in these children the examination accomplished was so inadequate as to be of no value in assessing whether sensory defect was or was not present.

The terminology and classification of cerebral palsy in childhood.

The scope of cerebral palsy.

Cerebral palsy is a descriptive term applied to a group of chronic conditions occurring in childhood in which paresis, inco-ordination or involuntary movement due to cerebral lesions prevent full use of the limbs. Tacit agreement appears to have been reached that conditions characterised by a progressive course and those of an acute and transitory nature should be excluded from the scope of the term cerebral palsy. At the same time what should and should not be classified as cerebral palsy has been arbitrarily defined and varies in different centres, as the importance of the many aspects of the condition are differently stressed.

History.

Originally what was understood as cerebral palsy was a collection of conditions showing stiffness or other abnormality of limb movement, due to cerebral lesions following prenatal disease or injury at birth, (Cazauvielh, 1827, Little 1862).

As research into these conditions proceeded it became apparent that a number of them, in particular hemiplegia, resulted from post-natal conditions as well as from abnormal parturition as emphasised by Little. (Charcot, Marie 1885,1886, Strumpell, 1884).

Hemiplegia /

Hemiplegia resulting from acquired disease in childhood was therefore admitted into the scope of cerebral palsy. But at the same time the groups described by Little as generalised or paraplegic rigidities tended to be excluded as it was considered that they were most commonly of spinal origin, - a form of childhood tabes dorsalis characterised by rigidity or spasticity. (Dejerine,1903, Erb, 1877, Brissaud 1896).

Other authors considered that these conditions were cerebral, not spinal in origin, (Heine, 1860, Parrott, 1873, Freud 1893). When this point of view became more prevalent the conditions, termed diplegia by Freud, were readmitted into the category of cerebral paralysis.

At the end of last century the scope of paralysis had been enlarged to include conditions of disorder of movement, which were believed by Freud to be related to his group of diplegic disorders, and were included in that term. (Freud, 1893, Ganghofner, 1895, Haushalter, 1895).

The classification at that time was summarised by Freud, (1893) as follows;

Hemiplegia. /

Hemiplegia.

Diplegia.

Generalised rigidity.

Paraplegic rigidity.

Bilateral hemiplegia.

Generalised chorea and bilateral athetosis.

At the same Freud and other authors were greatly troubled by the difficulties of defining the scope of cerebral palsy and the classification of the types of the condition they had recognised. In particular they were concerned with the relationship of epilepsy, mental defect and types of ataxia to cerebral palsy. In particular they were unable to differentiate between ataxic conditions resulting from birth injury and cases of the forms of progressive ataxia which had recently been described as occurring in later childhood. (Friedreich, 1876, Marie, 1893, 1914). Since they believed that the relatively rare ataxic conditions which they encountered were progressive they excluded them from their categories. Only when it was realised that a number of ataxic conditions were not progressive were they included in the scope of cerebral palsy. Recent authors all include ataxia as a category and suggest, indeed, that the incidence of ataxia in a series is some indication of how efficient ascertainment of cases has been. (Phelps, 1941; Perlstein 1952; British Council for the Welfare of Spastics, 1948).

Classification of types of cerebral palsy in childhood./

Classification of types of cerebral palsy in childhood.

Classification of cerebral palsy today is by symptomatology or clinical picture. Since the attention paid to different manifestations of cerebral palsy varies from centre to centre, and since they are so diverse, it is inevitable that classifications should vary greatly.

Other attempts at classification.

Frequent attempts have been made to improve upon classification of types of cerebral palsy by symptomatology or clinical picture from time to time. Since the manifestations of cerebral palsy are so diverse in type and so numerous it has been argued that classification by symptoms and signs can only take account of a few symptoms and signs in any one case of cerebral palsy. Any classification based on these must inevitably lead to the grouping together of cases differing very greatly in their disabilities. The label given to them can only be very approximately a true description.

To remedy this a number of attempts have been made at different times to classify cases by other criteria, most commonly on a pathological or an aetiological basis.

The very first attempts to define the types of cerebral palsy were made by pathologists who were chiefly interested in types of cerebral atrophy occurring in children in the prenatal period and as a result of birth injury. Attempts were made to define the clinical

picture /

picture to be expected in each form of atrophy or agenisis described. Thus, as a result of the work of Kundrat, (1882) and Audry, (1888) on porencephaly many attempts were made to describe a clinical syndrome which could be regarded as being pathognomonic of cerebral porencephaly. (Brissaud 1896). As a result of the early work on cerebral atrophy by Cazauvielh,(1827), Cruvehlier, (1862) and Henoch (1842), clinical criteria were suggested for the diagnosis of lobar sclerosis, (Cotard, 1868).

Though the possibility of classifying cases according to cerebral pathology have obviously much improved with the development of recent special radiographic techniques, the criticisms of attempts at a pathological classification made by Freud ramain valid. It was difficult to define the extent of cerebral lesions clinically. In porencephaly, for example, the extent of actual cyst formation gave a very poor indication of the actual degree of cerebral disturbance that was present. In many cases the cerebral lesions were as diverse in type as their resulting manifestations, and where manifestations could be observed in detail, cerebral pathology could not. Freud looked forward to the day when it would be possible, because of greater knowledge of cerebral localisation, to diagnose the site of lesions in the brain more accurately than in his day. To some extent his hopes in this direction have been fulfilled and with the increasing clarity achieved in the definition of the results of lesions in pyramidal, extrapyramidal, cortical /

cortical and subcortical regions more accuracy is now possible in clinical as well as direct radiographic and neurosurgical diagnosis. But the difficulties of assessing the importance of each of a large number of cerebral lesions occurring in patients with multiple neurological abnormalities, remains.

Attempts at aetiological classification were also made at a relatively early stage of the interest in cerebral palsy. Indeed, it can be argued that the title of Little's second paper on the subject of "The influence of abnormal parturition, difficult labours, premature birth and asphyxia neonatorum, on the mental and physical condition of the child, especially in relation to deformities", implied an aetiological classification. With the realisation that a large proportion of cases showing hemiplegia in childhood were due to postnatal causes and most cases of diplegia to prenatal or natal causes, it was clear that some attempt would be made to differentiate these cases more formally. It was suggested that prenatal factors, and especially premature birth were almost always responsible for paraplegic rigidity, for example, (Brissaud, 1894), and classifications were suggested which comprised three categories, taking account of those with prenatal, natal and postnatal aetiologies.

TABLE I. /

TABLE I.

The aetiological classification of cerebral palsy suggested by Sachs 1891.

Groups.	Clinical forms in order of frequency.
Paralysis of intra uterine onset.	Diplegia Paraplegia Hemiplegia
Birth palsies.	Diplegia Paraplegia Hemiplegia Diataxia Cerebellar form
Acute (acquired) palsies.	Hemiplegia Paraplegia Diplegia Choreic and athetoid disorders, unilateral and bilateral.

The most complete of these was that of Sachs, (1891) which is illustrated in Table I. It is of interest that the author himself later modified his suggestions for classification, admitted that the classification by aetiology was not exclusive and that much clinical variation occurred in the types of cases resulting from similar aetiological factors.

Though knowledge of the aetiology of cerebral palsy has greatly increased since the time of Freud, his objections to attempts at classification on the basis of causal factors remains. He noted that it could take no account of the multiplicity of contributing factors usually present in the aetiology of cases of cerebral palsy. He cited cases where there appeared to be hereditary predisposition, maternal abnormalities during pregnancy and natal and postnatal abnormalities, the relative importance of each being obscure. The liability of the child with hereditary syphilis to suffer from cerebral arterial occlusions as a result of birth injury or later infectious disease was one of his examples. One of the largest groups in any honest classification of cases of cerebral palsy based on aetiology would be under the heading of "Unknown". Moreover the results of similar aetiological factors varied very greatly in different patients, in whom the types of disorder, severity, course and complications encountered might be very /

very different.

With the recent stress laid by American authors, on the necessity for any classification of cerebral palsy to give information of value to the therapist, the use of aetiological classification has further diminished. (Phelps, 1941.)

Symptomatological classification.

Sy symptomatological classification is meant classification based on symptoms and signs shown by patients with cerebral palsy. It is this form of classification that is generally used today. Yet the variations of terminology that are encountered are thoroughly confusing and so diverse that the greatest care has to be exercised in the comparison of very similar looking series of cases. The variations of classification and terminology are due largely to differences in the outlook of workers in different schools and countries when the first attempts at classification were made. Many of the confusions in classification that are encountered, though thoroughly annoying are of great historical interest.

Though he did not define his terms accurately, Little used rigidity as a term to indicate cases in which impairment of movement appeared to be due to stiffness of a limb. Thus he wrote of hemiplegic, paraplegic and generalised rigidity. He also noted that movement could be disordered without the limb being stiff. Charcot was greatly troubled because he had found no

cases /

cases of rigid hemiplegia in 40 years neurological experience. Rigidity, for him was a condition of increased tone which might be contrasted to spasticity of hypotonia. To Little it merely meant stiffness and his cases were almost certainly congenital hemiplegias in whom the increase of tone was spastic. Osler classified cases similar to those described by Little, as bilateral hemiplegia largely because they showed spasticity, not rigidity on examination (Osler 1889). Little's description of paraplegic and generalised rigidity to describe other of his cases was generally accepted however and persisted until recent American classifications emphasised that in a number of such cases much of the increase of tone was of spastic type.

The occurrence of involuntary movements in association with volitional muscular activity had been noted by a number of early authors. Little 1843, 1862, Cotard 1868. The first full description of the slow writhing involuntary movements of the fingers and toes found in cases of hemiplegia was given by Hammond (1881). He attempted to define a disease entity on the basis of his findings, ignoring the fact that the movements he described were associated with other well known clinical syndromes in all his cases.

He termed his syndrome "Athetosis". It was soon pointed /

pointed out that the involuntary movements described by Hammond occurred, in fact, in a large proportion of children with hemiplegia, and in a smaller proportion of adult patients. Nevertheless great interest was aroused by Hammond's description and a large number of studies appeared in which athetosis was claimed to exist in large numbers of cases. In some of these papers a clear distinction was made between movements like those which had been described by Hammond and those of a more rapid, choreid nature. (Audry 1892, Raymond 1876). In others, however, no such distinction was made and the term athetosis was applied to involuntary movements of many types, occurring in various conditions (Bouchaud 1824). In spite of the superb analysis of the different types of involuntary movements, encountered in hemiplegia and other conditions, by Gowers (1876) the term athetosis was indiscriminately used by others to apply to slow or rapid movements occurring in the fingers and toes only or of generalised, unilateral or bilateral extent. Hence the present confusion; athetosis meaning one clinical condition in adult neurology and a number of different conditions in cerebral palsy in childhood arose. Even in recent studies of the latter condition athetosis may mean athetosis or be applied, in addition, to choreoid movements, choreoathetosis and dystonia indiscriminately.

<u>The basis of symptomatological classifications of cerebral palsy.</u> /

The basis of symptomatological classifications of cerebral palsy.

This form of classification is based on the manifestations of their cerebral lesions that may be observed in those who suffer from cerebral palsy. Since the manifestations are many and diverse in type any classification will be, at best, an approximate description of the disorder from which the patient suffers. Symptoms and signs of minor importance must be ignored. The particular manifestation of disorder selected as a basis for classification will vary from centre to centre according to the differing degrees of importance attached to different symptoms, and the orientation of the particular clinician concerned.

The traditional classification of early workers after the time of Little was by neurological syndromes. These will be discussed in more detail later. Recent American classification has tended to group cases on the basis of single clinical features of these syndromes. Thus rather than classify cases as hemiplegic or diplegic they are classified as atonic, spastic or rigid: rather than classify cases as suffering from post encephalitic Parkinsonism they are classified under "Tremor". Isolated clinical features rather than syndromes are the basis of classification (Phelps, 1940, 1943; Hellebrandt, 1951).

TABLE II. /

Recent American Classification of Cases of Cerebral Palsy.

TABLE II.

I. Spastic.
- A. Aspastic.
- B. Spastic.
  - (1) Monoplegia.
  - (2) Hemiplegia.
  - (3) Paraplegia.
  - (4) Triplegia.
  - (5) Quadriplegia.
- C. Basilar.

II. Athetosis.
- A. Tension.
- B. Non-Tension.
- C. Dystonic.
- D. Flail.
- E. Arm Neck.
- F. Deaf.
- G. Shudder.
- H. Hemi-Athetoid.
- I. Cerebellar Release.
- J. Rotary.
- K. Emotional Release.
- L. Tremor.
- M. Unclassified.
  - (1) Paraplegia.
  - (2) Quadriplegia.
  - (3) Monoplegia.
  - (4) Recovered.

III. Rigidity.
- A. Intermittent.
- B. Continuous.
- C. Miscellaneous.
  - (1) Hemiplegia.
  - (2) Paraplegia.
  - (3) Triplegia.
  - (4) Quadriplegia.

IV. Tremor.
- A. Intention.
- B. Constant.

V. Ataxia.
- A. Cerebellar.
- B. 8th. Nerve.

Table II. shows a recent widely accepted American classification of cerebral palsy based on that of Phelps. (Hellebrandt 1951.) Cases are divided into five groups and many sub-groups. The aim of the classification, and others like it is to give a brief description of the case which will be helpful to the therapist. In some measure it succeeds. In the group called athetoid, for example, classification by the various sub-groups allows nearly all the more commonly occurring types of involuntary associated movement to be described. The treatment, which varies greatly in the different types can be prescribed correctly on this basis. On the other hand the classification is much less successful in the achievement of its aim, of helping the therapist in other types of cerebral palsy. Under the heading of rigidity for example, in the sub-group, C. Miscellaneous, are found cases of hemiplegic, paraplegic, triplegic and quadriplegic rigidity. The hemiplegic cases show rigidity, presumably because of contracture, the paraplegic, triplegic and quadriplegic cases show rigidity predominantly because of imbalance in the neurological mechanisms controlling muscle tone. The treatment of the rigidities due to such different causes are necessarily markedly dissimilar.

Objection may also be taken to the classification on the /

the grounds of the choice of the manifestations on which it is based, and because of some of the inherent contradictions in it. Thus spastic and rigid conditions are considered as very different categories, yet alteration of muscle tone is one of the most difficult findings to assess. Alteration in muscle tone is variable at different times and in different conditions and positions in many patients. Very frequently in hemiplegic, paraplegic, triplegic and quadriplegic patients spasticity and rigidity co-exist. Indeed in the symmetrical disorders a mixture of rigidity and spasticity is the rule rather than the exception. Moreover children showing aspastic spasticity, Group I, Sub-group A, must be excessively rare, and if the terms are to be accepted in their usual sense cannot exist at all.

The test of this and similar classifications comes when cases are selected and grouped for discussion of their aetiological findings.

TABLE III./

TABLE III.

Classification of Perlstein. 1952

By Clinical Symptoms	Topographical Involvement of extremities.	By Muscle Tone.	Severity.	Etiology.
Spastic conditions	Paraplegia	Istonic	Mild	A. Prenatal I. Hereditary (a) Static (b) Progressive
	Diplegia			
Dyskinesias	Quadriplegia or Tetraplegia	Hypertonic	Moderate	2. Acquired in Utero (a) Infection (b) Anoxia (c) Cerebral haemorrhage (d) Rh. factor (e) Metabolic disturbance (f) Gonadal irradiation
Choreas	Hemiplegia	Hypotonic	Severe	
Athetoids	Triplegia			
Dystonia	Monoplegia			B. Natal Factors. I. Anoxia 2. Cerebral haemorrhage (a) Trauma (b) Pressure change etc.
Tremors	Double Himiplegia			
Rigidity	Limited to both Upper extremities			C. Postnatal Factors I. Trauma 2. Infections 3. Toxic causes 4. Vascular accident 5. Anoxia 6. Neoplasms and developmental defects.
Ataxia				

Using this type of classification one author found it necessary, when wishing to discuss the aetiology of bilateral spastic conditions, to remove his sub-group of hemiplegic patients from consideration. He apparently felt justified in doing this because he recognised that cases of spastic hemiplegia were fundamentally different from cases of bilateral spastic disorder. Yet a classification which entails the discussion of such groups as I, leaving out sub-groups A, aspastic, B,(1) monoplegia (2) hemiplegia and C. basilar can hardly be considered sound. It would seem more rational to take the drastic step of resorting to the well tried historical classification of cases of cerebral palsy on the basis of neurological syndromes. At least by doing so such nosological confusions as classifying cases as Spastic, Aspastic, Spastic spastic, and spastic basilar, might be avoided.

The classification of Perlstein (1952) shown in Table III. is a very interesting and partially successful attempt to classify cases on the basis of a number of clinical findings, on the severity of the disorder and on aetiology. It recognises the fact that some indication is desirable of the severity of the condition, its extent and type, in any child with cerebral palsy. One of its great advantages is its relative flexibility. The classification can be used to describe almost any case of cerebral palsy with some accuracy. It has two severe defects however. It is clumsy and is not very

suitable /

suitable when series of similar cases are to be compared. The clumsiness means that though a case may be relatively fully described by the nomenclature of the classification, this frequently becomes but slightly less complex than a full case note. To comprehend what is implied by calling a patient a case of "Quadriplegic tension athetoid of mild degree due to anoxia at the time of birth" is something that requires more than a moment's thought. The other objection is that the informality of the classification tends to make it formless. It is difficult, therefore, to know which of a large number of possible descriptions contained in the classification to use as a basis for the systematic grouping of similar cases of cerebral palsy.

Nevertheless the classification is intelligible to clinicians who classify cases by aetiology, by neurological syndrome or by isolated clinical findings and there is a great deal to be said for the rather full description of the types of cerebral palsy it permits.

Classification by neurological syndromes.

Classification by neurological syndrome was the method used by most of the early authors and though the details of classification varied they usually included two main categories, unilateral paralysis, consisting of hemiplegic cases and bilateral paralysis which included paraplegic and generalised rigidities, bilateral hemiplegia /

hemiplegia and usually, though not always, conditions in which disorder of movement was more prominent than actual paresis. Charcot (1887), Heine (1860), Bernhardt (1885), Gowers (1888.) The fullest of the early classifications was that of Freud which was generally accepted by his contemporaries, Seeligmuller (1879), Ganghofner (1895), Haushalter (1895) Sachs (1926). The classification is shown in Table IV. It will be seen that there were two main categories, unilateral disorders including hemiplegic disorder and diplegia including bilateral disorders. The latter comprised paraplegic triplegic and tetraplegic rigidity, bilateral hemiplegia and cases showing disorder of movement as their main symptom.

TABLE IV.

Classification of Freud, 1896

Cases of Cerebral Palsy in Childhood.

Unilateral disorders.

Hemiplegia.
right.
Left.

Bilateral disorders.

Diplegia.
Paraplegic rigidity.
Generalised rigidity.
Bilateral hemiplegia.
Choreoathetosis.

Possibly other groups.

It is on Freud's classification that the classification used in this survey is based. Table V. It will be seen that certain modifications have been made. The categories have been increased in number to include patients suffering from ataxia. This becomes necessary since it is now generally agreed that though not strictly cerebral palsies, cases of ataxia of non progressive type should be included in the class of cerebral palsy. As a result of experimental and pathological work done since the ti e of Freud it has become increasingly clear that many of the cases showing disorder of movement rather than paresis as their predominant symptom are not related to cases of diplegia as closely as was thought. Sherrington (1947), Magnus (1926) Walshe (1923). It becomes necessary, therefore, to make a special category for these cases and it is better called dyskinesia than by any other term, which is also applicable to specific types of involuntary movement (Perlstein, 1952).

Double hemiplegia is also removed from the category of diplegia. It was included under this heading by Freud in order to achieve some uniformity with American classification, which as a result of the work of Osler had called cases of generalised rigidity bilateral hemiplegia. (Osler 1889, Lovett 1888). Since the cases /

cases of bilateral hemiplegia differ from diplegic cases in the greater severity of the involvement of the arms than the legs and since they differ from cases of unilateral hemiplegia in the severe involvement evident in the cranial nerves it has been felt better to make a special category of them.

Similarly because the aetiological findings, course and findings on examination of cases of diplegia with ataxia and diplegia without ataxia differ a separate category has been made for these cases. In this category are cases similar to those called diataxia by Ramsay Hunt (1918) Sachs and Haïsman (1926).

TABLE V. /

TABLE V.

Classification of Cases of Cerebral Palsy in Childhood.

Neurological diagnosis.	Extent.	Severity.
Hemiplegia	(Right (Left	(Mild (Moderately severe (Severe
Double hemiplegia		(Mild (Moderately severe Severe
Diplegia Hypotonic) Dystonic) Rigid or spastic)	(Paraplegic (Triplegic (Tetraplegic	(Mild (Moderately severe (Severe
Ataxic diplegia Hypotonic) Spastic) With contracture)	(Paraplegic (Triplegic (Tetraplegic	(Mild (Moderately severe (Severe
Ataxia Cerebellar) Vestibular)	(Predomin- (antly (unilateral (Bilateral	(Mild (Moderately severe (Severe
Dyskinesia Dystonic) Choreoid) Athetoid) Tension) Tremor)	(Monoplegic (Hemiplegia (Triplegic (Tetraplegic	(Mild (Moderately severe (Severe
Other.		

The categories of cerebral palsy in the present classification.

In this section it is proposed to define the categories in the classification more clearly and to describe briefly, on what basis cases are grouped in each.

Hemiplegia. The clinical manifestations of cases with hemiplegia resulting from prenatal or natal conditions and those resulting from postnatal factors were found not to differ significantly and no attempt is made to distinguish them in the classification.

The side is noted because the disability tends to be greater if the master hand, usually the right, is involved. Speech defects and aphasia are also commoner in those with right hemiplegia than in those with left hemiplegia.

The severity is assessed on the basis of the degree of impairment of function in the affected arm. It is true that an intelligent patient with good attentive parents may make much more use of a severely affected limb than a mentally defective child in poor surroundings, but in general, if the degree of useful function is considered it is possible to place the majority of cases in one of three groups without much difficulty. Mild hemiplegias are those in which the affected arm is capable of being use independently and usefully. Moderately severe cases are those in which the affected arm /

arm is of use in assisting the unaffected arm but is not capable of independent useful function. Severe cases are those in which the upper limb is so impaired that for practical purposes, useful function is absent.

Though it is true that most of the severe cases show very severe contracture and it is therefore tempting to classify cases of hemiplegia in to those with contracture and those without. The objection to this is that in hemiplegia in childhood some contracture is almost always present even in mild cases, and in a small proportion in all groups, athetosis of the fingers and distal parts of the upper limb, and in these contracture occurs tends to be less marked. (Freud 1897)

The classification cannot take account mental deficiency, epilepsy, aphasia, speech defects, athetosis of the fingers, or hemianopia as it would become too large and complicated if sub-groups for those with these disorders were included.

Double hemiplegia.

Cases classified as suffering from double hemiplegia show signs similar to those found in unilateral hemiplegia bilaterally. But in addition they show some degree of involvement of the trunk muscles, and cranial nerves. A large proportion appear to be of congenital origin and show mental deficiency, small skulls, high palates and other skeletal abnormalities. Many are epileptic.

The /

The cases are distinguished from cases of diplegia by the greater involvement of the arms than the legs which they show, by the more or less severe involvement of the trunk muscles and cranial nerves and by the different course of the condition. Few of those showing double hemiplegia are born prematurely and they never show the classical stages of diplegia, especially the dystonic stage.

Cases of double hemiplegia do not show the contrast between the relatively well developed shoulder and upper trunk and poorly developed pelvis and legs which is so commonly found in diplegia. On the other hand the tendency to contracture in the limbs is even more marked than in diplegic cases.

Because such a high proportion of patients with bilateral hemiplegia show bulbar involvement there is always severe speech defect and owing to the liability to inhalation of food matter respiratory infections, and death are common.

Double hemiplegia is always a severe condition but there are differing degrees of functional disability. Mild cases are those with some useful function of the upper limbs, moderately severe cases with useful function in the lower limbs, if not in the upper. Severe cases are those who are bedridden on account of their paresis and the limbs incapable of useful function.

Diplegia. /

Diplegia.

By diplegia is understood a condition of more or less symmetrical paresis of the limbs more severe in the legs than the arms and dating from birth or shortly after. Commonly, but not necessarily the paresis is accompanied by rigid or spastic increase in muscle tone. In this classification diplegia complicated by cerebrellar ataxia is classified separately from diplegia without this finding, for reasons which will be considered in detail later.

The category of diplegia is subdivided into four groups of cases according to the stage of the condition which the child demonstrates. It has been found that the histories of diplegic patients show remarkable similarities. In the histories, 3 clear-cut stages may be recognised in a large proportion of cases.

Initially most cases are born without apparent gross abnormality but frequently when a detailed history is obtained the mother admits that in the first weeks after birth the child was floppy and showed poverty of limb movement. When the child was picked up "The head rolled all over the place doctor, when he was laid down and wrapped up he stayed put till morning so that you would think he hadn't moved all night." On examination during the first weeks after birth the child is hypotonic on examination, and this finding of hyptotonicity is frequent enough /

enough for the first stage of diplegia to be characterised as the hypotonic stage.

At the age of 6 to 16 weeks in those with approximately normal milestones a change in the child is noted by the mother. When his position is changed abruptly, most commonly when he is being fed or bathed, the child suddenly goes stiff in her arms. He is stiff, "all of a piece", the head is thrown backwards, the back is arched, the arms held stiffly by the sides and the legs extended with the feet in a position of equinus. After a few seconds the child appears to be normal again. On examination during this stage abrupt extension of the head will produce a position of the baby similar to that described by the mother. The child is hyperextended, the back is arched, the arms are adducted at the shoulders, extended at the elbows, ronated at the forearms and flexed at the wrists and fingers. This position, similar to that of decerebrate rigidity in many respects, is produced by means of a rather sudden dystonic movement of the trunk and limbs on stimulation. It is produced on extension of the neck or trunk in an abrupt manner initially, but as the weeks pass may be produced more and more easily by change in the position of the child. After 2-3 weeks it is sometimes possible to produce it by merely banging on the sides of the cot. When the child is observed during these dystonic attacks consciousness seems to be impaired to some degree in a proportion of /

of cases but in none in the present series was it lost.

The stage of dystonia, or dystonic movements, occurs first when the child is beginning to hold up the head and persists until postural mechanisms are advanced enough for him to be able to retain the sitting position with a little support and to use the hands voluntarily in a more or less intelligent fashion. This co-incides in all cases in this series with the stage of being able to maintain the head in a position of slight flexion. The dystonic movements are very similar to those demonstrated experimentally in decerebrate dogs, and termed extensor thrust by Sherrington (1905). It is tempting to suggest that they are the result of postural reflexes unhibited by cortical centres, which have been damaged or are immature.

After 6 to 12 weeks in those with approximately normal milestones the dystonic attacks become less frequent. They are more difficult to produce, and frequently the mother finds that they occur only when the head is allowed to drop suddenly as she places the child horizontally prior to putting him in bed. As the attacks decrease in number she notices another change in her child. In contrast to his floppy state after birth she finds that he is beginning to be stiff when handled. The stiffness is commonly noted first in the thighs. The mother notes that she has to separate the thighs /

thighs with force in order to place the child's napkin, or she may pass it between the legs on the end of a pencil. In other cases she finds that one or both hands have to be opened in order that they may be washed, the the fingers are stiff when she does this.

This stiffness of the hands and legs on handling indicates that the next stage of diplegia is beginning, the rigid or spastic stage. As the dystonic attacks cease the rigidity of the limbs, usually more marked in the legs than the arms, increases. By the time the attacks have ceased completely there is considerable rigidity in the legs and appreciable rigidity in the arms. At about this stage the child is beginning to sit, first with support and then without it; he is using the hands to play with toys in an increasingly intelligent manner; he is maintaining the head in a position of slight flexion, apparently in order to see more clearly what he is doing with the arms. In those with normal milestones the dystonic attacks disappear at the age of between 6 and 8 months, at the time when voluntary activity is becoming manifest.

When the rigid or spastic stage become evident with increase of tone in the legs and arms it is initially associated with a tendency to involuntary extensor movements in the limbs and at this stage of its development /

ment the tendon jerks are diminished rather than increased. At the age of 10 months to 12 months in those with normal milestones a change occurs however and the tone, hitherto predominantly rigid becomes spastic in type, the tendon jerks increase and the Babinski responses positive. The limbs being to take up flexor, rather than extensor, positions; they may show athetosiesis and develop contractures; the manifestations are rather similar to those of adult pyramidal lesions from this time onwards.

The development of contracture in the limbs is the almost invariable end result of the rigid and spastic stage of diplegia. Once it appears contracture tends to be progressive and extremely difficult to treat by surgical, physiotherapeutic or other methods.

The distribution of paresis in the limbs in diplegia was considered more from the functional point of view than from the neurological. Neurologically extremely few cases showed abnormalities confined only to the lower limbs; in the majority neurological abnormalities of relatively slight degree were still functionally useful. In this series the functional impairment of the limbs was the criterion of classifying the extent of involvement. Thus paraplegic cases were those with the lower limbs affected but both upper limbs functionally useful, whether they showed neurological abnormality or not.

The severity of diplegia was again assessed on the basis /

basis of the degree of impairment of function. In general this corresponded with the degree of neurological impairment which was evident, but not in all cases. Severe cases were those in which it was felt that walking without support would be impossible except for very short distances. In those classified as moderately severe, walking was possible without support but the gait was impaired. Mild cases were those in which walking without support was possible and no gross abnormality except clumsiness or perhaps some slight unsteadiness was present.

By describing cases of diplegia on the basis of the stage of the condition which the case shows, the numbers of the limbs affected and the severity of functional impairment, a fairly comprehensive description of the patient's disorder is achieved.

Ataxic diplegia.

A group of cases was found in which diplegia was associated with cerebellar ataxia. The number of cases included in this group was smaller than that in the group of diplegia without ataxia. The cases in the groups differed in aetiology, course, findings and complications. A family history of neurological abnormality, epilepsy or mental deficiency was often obtained. Premature birth, abnormality of pregnancy and neonatal asphyxia were less frequently present aetiological /

aetiological factors in those with ataxia than in those without. Mental deficiency was present in a higher proportion of cases of ataxic diplegia and was frequently the presenting symptom. The cases showed no dystonic stage and spasticity, uncomplicated by rigidity appeared during the hypotonic stage in most cases. The children tended to have small heads or to be hydrocephalic. Scissoring of the legs, and extensor positions of the arms on movement were never encountered. Involuntary movements were flexor in type and the gait was marked by unsteadiness, deviation to one side or the other when blindfold, and a broad based position of the legs.

In addition to paresis, associated movements and contractures the limbs also showed a degree of inco-ordination of movement in patients suffering from diplegia with ataxia. Rapid and fine movements were especially severely impaired and generally more markedly on one side than the other. Pass pointing could be demonstrated in the limbs of most cases.

It was felt that cases showing the combination of ataxia and spastic diplegia occurred frequently enough, and differed markedly enough from those without ataxia to justify their inclusion in a separate category from these. Similar cases were described under the heading of diataxia by Ramsay Hunt (1918). They were classified by /

by stages, hyptotonic or spastic, and by the extent and severity of their limbs involvement in a similar manner to patients with diplegia who did not show ataxia.

Ataxia.

Cases in which the predominant finding was ataxia were grouped according to whether their ataxia was cerebellar or vestibular in type. They were also grouped according to whether they showed ataxia which was apparently symmetrical in its severity or ataxia which was predominantly unilateral. The severity of the ataxia was assessed on the basis of the functional impairment evident in the child's ability to perform normal tasks. Mild cases were those in which the child's activities were not limited; moderately severe cases were those in which some normal activities were rendered impossible and severe cases were those in which the performance of even simple tasks was much impaired.

Dyskinesis.

This category includes cases in which involuntary rather than actual paresis is the presenting manifestation. The groups included differ in aetiology, course and clinical findings.

It is probable that it will be possible with increasing study of forms of dyskinesis, to distinguish the types more accurately, and define clinical entities for /

for them. Thus it has been found possible in the present study to demonstrate the very close connection some cases with dystonia have to diplegic paresis. At present however it has not been found possible to define other types of abnormal involuntary movement accurately enough for their more accurate classification.

One is left with a heterogeneous group of cases with various types of involuntary movement as their presenting symptom. The types of involuntary movements encountered have been classified somewhat arbitrarily into dystonic, implying abnormal movement of the trunk as well as the limbs, choreoid, athetoid, tensor and tremulous. Choreiod movements are those in which the involuntary movement of the limbs is quick and maximum in the proximal, rather than the distal joints. Athetoid movements are those which are move evident in the distal than the proximal joints, and are slow and writhing in character. Tremor is usually evident in dyskinetic patients only on voluntary movement or in conditions of emotional stress. It is usually maximal in the distal joints of the limbs and may be associated with the two previous conditions. Tension is the description given to checking and limitation of voluntary movement which results from a sudden generalised increase in muscle tone when attempts at voluntary movement is made. Unfortunately, since these categories /

categories are not mutually exclusive, the classification of types of dyskinesis has to be based on a number of the presenting manifestations. Thus many cases of choreoid movement show a marked generalised increase of muscle tone when voluntary movement is attempted. Classification has to take account of these multiple forms of involuntary movement and activity.

The number of limbs involved in dyskinesia is indicated by classifying cases as monoplegic, hemiplegic, paraplegic, triplegic and tetraplegic. Trunk involvement is indicated by the phrase dystonic appearing in the description of the type of involuntary movement exhibited. On the other hand it was not possible conveniently to indicate those cases in which facial and tongue involvement was present.

Mild cases are those in which normal activity is possible with the affected limbs in spite of the abnormal movements. Moderately severe cases are those in which normal activities are restricted because of the abnormal movements, severe cases are those in which useful movement of the limb is virtually impossible on account of involuntary movement.

Other categories.

A few less frequently occurring syndromes were classified in this category. Thus cases of postencephalitic Parkinsonism, the mixed neurological pictures resulting /

resulting from other rare forms of encephalitis and cases in which the diagnosis is in doubt are better classified under the heading of 'Other' than incorrectly in some more accurately defined category.

In this survey it was found necessary to classify only 2 cases in the category of 'Other' syndromes. Other cases which might, with some justification, have been included, were in fact left out of the series of cases to be considered, as they were not considered, strictly, to be suffering from cerebral palsy.

Description of the clinical findings in 208 cases of cerebral palsy examined during the Survey.

In this section the results of the examination of the patients suffering from various types of cerebral palsy in childhood, will be described. The numbers of patients in the various categories are shown in Table LV The table, though important in showing the incidence of the types of cerebral palsy to be discussed, in fact, gives very little real indication of the importance of cerebral palsy in the community. It gives no indication of the severity of the disabilities from which the patient suffers and can take no account of manifestations of cerebral damage other than actual paresis of limbs. Thus though specific forms of educational difficulty are very common in various forms of cerebral palsy, until more is known about these it is not possible to assess the size of the burden which cerebral palsy presents to the educational system. In order that disabilities associated with the actual limb paresis may be considered it is necessary to discuss the clinical findings in patients in each category in some detail. Only then can the diversity and importance of all the manifestations of cerebral damage be assessed.

Hemiplegia.

75 cases were classified as suffering from hemiplegia by which was understood a paresis affecting the limbs of one side of the body, of cerebral origin and generally more severe in the arm than the leg.

In 33 patients there seemed to be little doubt that the hemiplegia was acquired as a result of postnatal disease, in 30 that it was due to abnormality of pregnancy or delivery or neonatal asphyxia and in 12 the cause was uncertain but thought to be congenital in most. Though the clinical impression was that acquired hemiplegia was, in general more severe, and attended by more associated abnormalities than the congenital variety, a careful examination of the results obtained during the survey failed to substantiate this. The sex ratio, side of the paresis, the severity of the paresis and the associated disabilities resulting from the different aetiological factors are shown in Table VI . It will be seen that in no instance except in sex ratio and aphasia was the incidence of abnormality different in the congenital and acquired groups, and even in aphasia the numbers are too small for the differences to be of statistical significance.

Sex.

In this series males outnumbered females by more than /

TABLE VI

COMPARISON OF THE FINDINGS IN CONGENITAL AND ACQUIRED HEMIPLEGIA IN CHILDHOOD.

	Male	Female	Rt.	Left	Totals	Severity of hemiplegia			More than 1" shortening in affected upper limb	Marked Athetosis	Severe Trophic Changes	Epilepsy	Asphasia
						Mild	Moderately Severe	Severe					
Congenital	20	10	20	10	30	11	8	11)	15	11	7	9	3
Probably Congenital	9	3	7	5	12	5	5	2)					
Acquired	23	10	17	16	33	11	13	9	11	8	5	15	9
Totals	52	23	44	31		27	26	22	26	19	12	24	12

than 2 to 1, in contrast to the figures obtained in most series, which tend to show an equal sex distribution of hemiplegia. (Osler, 1889; Lovett, 1888; Freud and Rie 1891). One possible explanation of the apparent discrepancy is that the older series tended to include a greater proportion of acquired cases of hemiplegia with a dramatic change from the healthy to the crippled child. This survey was more representative in that it included both, cases with dramatic histories, the acquired variety, and less dramatic cases with obscure aetiology, and frequently relatively mild paresis. The figures obtained during the survey tend to show a more equal incidence of hemiplegia between the sexes in the acquired than in the congenital form of hemiplegia. It is interesting to note that at least two recent surveys of the incidence of cerebral palsy show a higher incidence of hemiplegia in the male than the female, especially in congenital cases. (Asher and Schonell, 1950; Keddie, 1946). One earlier survey reported that hemiplegia was commoner in males. (Sachs and Peterson 1890).

Side of the paresis.

The side of the paresis is of importance from two points of view. Firstly, especially in acquired cases a change from the dominant hand tends to make a comparable degree of paralysis a more severe handicap than is the disuse of the less dominant hand. Adaptation to the disability is more difficult. Secondly when the master

side is affected, speech difficulties, aphasia and certain reading and writing handicaps are more frequent than when the non-dominant side is affected.

Table VII shows that whereas the severity of the paresis did not appear to depend greatly on the side of the body affected by hemiplegia, certain of the associate abnormalities were more frequent in right than left hemiplegia. It is interesting to note that only one case was encountered of aphasia occurring in a child with acquired left hemiplegia and he was left handed before the onset of his disease.

Throughout the series one was struck by the apparent ease with which children adapted to the paresis of one side, and the rapidity with which they learnt to do quite intricate tasks with the unimpaired hand. The reason why the paresis of the right side seemed to be a greater handicap, in some patients, than paresis of the left were that so many social and educational techniques are designed for the right rather than for the left handed.

In most previous surveys hemiplegia affects the sides apparently equally frequently, or the right rather oftener than the left. (Osler, 1889; Freud and Rie 1891 Lovett, 1888).

<u>The severity of hemiplegia</u>.

The severity of the hemiplegia is of far more importance /

TABLE VII

The findings in 75 cases of hemiplegia by sex and the side of the body affected.

		Severity					Schooling								
		Mild	Mod.	Severe	Total	Epil-epsy.	Normal	P.H.	M.H.	Ineducable	Not yet at school	Aphasia	Speech defect	Visual defect	Overactive behaviour
Male (Rt.	31	12	10	9)		8	7	5	5	5	9	11	9	6	4
Male (L.	21	8	8	5)	52	6	9	5	4	1	2	1	5	1	3
Female (Rt.	13	4	4	5)		5	3	2	2	2	4	0	4	1	2
Female (L.	10	3	4	3)	23	5	4	2	1	1	2	0	3	3	0
	75	27	26	22	75	24	23	14	12	9 ·	17	12	21	11	9

P.H. = Physically handicapped.
M.H. = Mentally handicapped.
Mod. = Moderate.

importance to the child than the side on which it occurs in the majority of cases. In this series cases have been grouped according to the severity of the loss of function they show in the affected arm. Mild cases were those in which the upper limb, and especially the hand, was capable of independent voluntary activity. In general children with mild hemiplegia used the hand somewhat clumsily; fine and rapid movements of the fingers were impaired. Restriction of voluntary movement was usually confined to some restriction of supination of the affected forearm and extension of the elbow. The tone of the limb was usually moderately, rather than severely increased and contracture when present, was slight. Limb shortening was not marked. Associated movement, the affected arm assuming a position of adduction at the shoulder, flexion at the elbow, wrist and fingers and pronation of the forearm were nearly always evident when the other arm or when the legs were used however. The tonic neck reflex was present though not usually striking. The Hoffmann sign might or might not be positive. The plantar response was almost invariably extensor.

Moderately severe cases used the affected arm alone only when they had to, either because they were made to or because the unaffected arm was already engaged. The everyday functions of the affected arm were confined to assisting /

assisting the normal arm. Handling was clumsier than in the mild group. Limitation of movement, contracture, increase of tone, associated movements, and especially the tonic neck reflex, were more marked. Involuntary movements of the fingers and vaso-motor disturbances, which tended to be relatively slight in the mild group were much more severe in the moderately severe cases. Sensory disturbances were sometimes encountered.

Severe cases were those in which the upper limb was so severely affected that, for practical purposes, the function of the limb was confined to giving some support to the unaffected limb and to objects on which the unaffected limb was operating. In many cases in the severely affected group the affected limb was incapable even of these very limited activities. About half the severely affected cases showed marked athetosis of the fingers on any attempt at voluntary movement. The paresis of movement, contracture, increase of tone, limb shortening and vaso-motor disturbances tended to be more severe than in the other groups and very frequently surgical operations had been necessary to render walking possible.

In the severely affected cases associated movements of the affected limbs were very marked and the tonic neck reflex strongly positive, though somewhat limited by contracture in some patients. To many of these children /

children their affected upper limb was more of an hindrance than a help.

The loss of function sustained in the affected limbs by children with hemiplegia is due to disorganisation of voluntary movement shown by the limbs themselves and to the inability, because of other disabilities, of the children to use the limbs as well as an otherwise normal child could. Thus a child with hemianopia tends to use an affected arm more clumsily than a child with a similar degree of paresis who has normal vision. Though it is realised that in many cases defects of the limbs cannot be distinguished from defects of the child, they will be considered separately in the following sections in order to make their discussion easier.

The relative severity of the paresis in arm and leg.

In general the paresis of the upper and lower limbs was proportionate, the arm more affected than the leg, but moderately affected in cases in which the paresis of the upper limb was classed as moderate and severe where it was classed as severe. This was not true in all cases however and in some the severity of the leg involvement seemed to be much less, or much more severe than might have been expected from the involvement of the upper limb.

Case II. Male Born 1938.

Moderately severe left hemiplegia dating from birth.

The /

The first of three children born to healthy mother aged 24 and her healthy husband. She had had a previous miscarriage. The second and third children were born after normal pregnancy and labour and are well.

She was well in her pregnancy with the patient and the delivery was spontaneous, in hospital at term, after a normal labour of 10 hours 15 minutes. The child appeared to be normal at birth, birth weight 8½ lbs. and the neonatal period was normal.

He held up his head at the age of 2 months and sat with support at the age of 6 months. He sat without support at the age of 8 months, stood without support at 1 year and walked very soon afterwards.

When he began to walk at the age of 1 year there was no abnormal position of the arms, (confirmed by photographs) but by the age of 15 months the left arm was flexed across the abdomen, seemed stiff on handling and he was limping. He began to walk on the toe of the left leg and dragged it as he proceeded.

Initially he seemed to handle toys normally, at the age of about 5 months but, again at the age of 15 months he began to "forget to use the left arm". There was no evident illness during infancy or between the age of 12 and 15 months. He said his first words at the age of 1 year and was saying sentences by the age of 2.

Because of his abnormal walking and an increasing tendency for the left arm to be cramped up across the chest with the hand clenched and the thumb underneath the fingers, he received physiotherapy from the age of 2. At the age of 4 he began to use the left arm to assist the right and could dress himself.

At the age of 5 he went to a normal school and did quite well there, always being in the top half of the class and able to play with children of his own age. I.Q. 110. In behaviour he tends to be quiet and tolerant but quite energetic and persistent when interested in what he is doing or wanting something.

Examination. Height 66½". H.C. 21⅞". There was 1½" shortening in the left arm and ½" of this was in the hand itself, compared to the right. The left leg showed ¾" shortening compared to the right. Adolescent.

He was intelligent, accurate in answers to questions and in responses to commands. Of good address.

The general muscular development was very good. There was a moderate degree of left facial paresis of upper /

upper motor neurone type. The power and co-ordination of the left side was impaired compared to the right. The impairment was only very slightly more marked in the arm than the leg. Voluntary and passive supination of the left arm was much limited, as were extension of the elbow, wrist and fingers. He was able to grasp objects with the left hand, but rather uncertainly and the grasp was rather poor. Rapid and fine finger movements on the left side were very poor but he was able to use them as well as one could expect. On any movement of the hand slow extension and athetosis of the fingers was evident and was utilised when he grasped. In the leg corsiflexion of the left foot was limited to the bare right angle and toe movements were impossible. When he walked he dipped to the left and there was a position of equinus of the left foot, slight flexion at the left knee and hip. He tended to swing the leg and put the foot to the ground heavily. When he ran he dragged the left leg. When he walked or used the left leg the moderately contracted position of the left arm became more pronounced with adduction at the shoulder, flexion at the wrist and fingers and pronation of the forearm. His gait was extremely clumsy and the movements of his left leg much more impaired than would be expected judging from the severity of the paresis in his arm.

The tone of the left arm and leg was spastic and increased compared to the right. The biceps, triceps, supinator, knee and ankle jerks were increased on the left though there was no clonus. The left Hoffman sign was positive on the left, negative on the right. The right plantar response was flexor, the left extensor. Sensory examination revealed no definite abnormality, no cutaneous or deep sensation. No Rombergism could be elicited. The left hand and foot were considerably colder than the right.

In only one case did the severity of limb involvement appear to be more marked absolutely as well as relatively in the lower limb than the upper.

Case 175. Male Born 1944

Mild left hemiplegia more marked in the leg than the arm following vague illness in infancy.

The first of three children born to healthy right handed mother. Other children normal pregnancies and births /

births and are healthy.

She was well in her first pregnancy with the patient and he was delivered at term after a normal labour lasting only a few hours, spontaneously in hospital. The mother had a post partum haemorrhage after delivery. The child was well at birth and cried at once. Birth weight was 6½ lbs. The neonatal course was normal and he sat with support at the age of 6 months and began to crawl at the age of 9 months. By this age he was playing a great deal with toys and using both hands.

At the age of about 9 months he began to be fretful and irritable. He slept poorly, seemed less ken on feeds and was lethargic. He crawled less and was less active. When he did crawl it was noted, after he had been off colour for about two weeks, that instead of crawling like a normal child, he dragged the left leg. The parents suspected infantile paralysis but a doctor could find no abnormality. After about 3 weeks of lethargy and fretfulness the child began to use the right arm much more than the left for playing with toys. At the same time his progress seemed to slow and he seemed in general to be less alert and interested in his surroundings than before. He began to be somewhat fevered at nights and failed to gain weight. These symptoms persisted until the age of 13 months and then gradually lessened, and fever ceased.

He stood at the age of 2 years and walked soon afterwards; when he did so the arm tended to flex across the chest, and he was very unsteady, falling a great deal. He gradually became steadier on his feet and from the age of 4 has been able to run and play normally with children of his own age. From about the same age he began to use the left arm, first to assist the right and then independently and freely.

He went to normal school at the age of 5 and has done moderately well there. The left hand has remained somewhat smaller and clumsier than the right.

He spoke his first words at about the age of 18 months and speech has developed normally with unimpaired pronunciation.

Examination. Height 49½". H.C. 20". The left leg shows ½" shortening and the arm ½" shortening compared to the right. He appeared to be normally intelligent answering questions readily, aggressively and intelligently and responding to commands briskly.

There was a latent weakness of the left arm compared to the right and the finger movements were slightly clumsier /

clumsier on the left. This was most evident on rapid and fine movements. There was slightly diminished power in the left arm compared to the right, but no limitation of voluntary movement could be detected apart from the impairment of movement of the fingers and the hand was used normally.

There was no limitation of movement of the left leg and dorsiflexion of the ankle, extension of the knee and abduction of the hip were considerably impaired in power compared to the right. There was defective co-ordination on the left. The left toes could not be moved voluntarily.

The gait showed dipping to the left and a tendency to walk on the left toes and to drag the left foot. There was some circumduction of the left leg.

The tone of the left arm and leg was somewhat increased and spastic. The increase was more marked in the leg than the arm and the biceps, triceps and supinator jerks though increased compared to the right were less increased than the knee and ankle jerks on the left in comparison. The Hoffman sign was absent on the right and probably positive on the left. The left plantar response was extensor, the right flexor. No clonus could be elicited.

When he ran or used the right arm powerfully there was a tendency for the left arm to assume the position of adduction at the shoulder, flexion at the elbow, wrist and fingers and pronated at the forearm. Extension of the elbow was readily achieved when the tonic neck reflex was used to elicit it.

No abnormal sensory findings could be detected. The left hand and foot were slightly colder than the hand and foot on the right.

In some cases involvement of the leg was relatively slight compared to that of the arm.

Case 100. Male Born 1945

Slight left hemiplegia. Mental defect. Mild ataxia. Congenital origin.

Born before marriage between healthy mother aged 20 at time of birth and father who was epileptic in childhood, as were two paternal uncles.

Second and third children were born after normal pregnancy and delivery and are well. During pregnancy was /

was well. At the start of labour, at term, she began to have profuse vaginal bleeding, which persisted and was admitted to hospital with the diagnosis of "Concealed and Revealed Accidental Haemorrhage". After 8 hours during which bleeding continued, she was delivered with episitomy and anaesthetic, of Stanley. There was considerable retroplacental clot but no infarcts noted. The birth weight was 6 lbs. 5 ozs. The child was very limp at birth and required oxygen and lobeline. Given hot bath. Wlow recovery. Thereafter normal neonatal course and was discharged at 10 days.

He was a windy regurgitating baby. Sat at 6 months with support and at about 10 months without. Walked at 18 months without support falling frequently and unsteadily, dragging his left foot.

Used the right hand from about 6 months but never the left and when he began to walk it was seen to be bent up across the chest.

Speech only began at 2½ and remained poor, slurred and difficult to understand. Attended school for the mentally handicapped and is a moderate pupil. I.Q. 65.

Examination. Height 47½". H.C. 20.6". 0.6" shortening of the left arm and .3" shortening of the left leg compared to the right.

Slow in thought and response. Speech defective in pronunciation and poor in content. Slight bilateral abducent paresis. Facial asymmetry is apparent on voluntary movement due to left sided lag, but is absent on rest and emotional movement.

Tongue movements clumsy and poorly co-ordinated but no definite paresis. Power of the left side was impaired and rapid and fine finger and toe movements were very poor. There was marked associated flexion of the left arm on any exertion or motor activity of the right arm. There was slight athetosis of the fingers on the left when he attempts movement with that hand. There is some limitation of voluntary wrist extension and of supination of the left forearm. Movements of the left lower limb are full.

The tone of the left side was increased in arm and leg. The increase is spastic in type and moderate in degree. The upper limb jerks are increased more, proportionately than the lower. The Woffmann and the Babinski were positive on the left.

Sensory. Superficial sensation, joint sense and vibration sense normal. There was a tendency to deviate to the right on blindfold walking, but no pass pointing.

Causes of the loss of function in the limbs in hemiplegia./

Causes of the loss of function in the limbs in hemiplegia.

The reason for the loss of function in the limbs affected by hemiplegic paresis are multiple and complex. As has been emphasised by recent experience in patients who have had hemispherectomy performed, they are little understood. (Knynauw, 1950). In the child the causes of the loss of function are complicated by the dwarfism and the more severe involuntary movements and trophic disturbances which tend to occur in the adult patient.

The child's inability to move the affected limbs as he wants to is due to damage to the voluntary motor centres, in part, and in part is due to other related and associated abnormalities of the involved limbs. The part played by the different factors in causing loss of limb function is worth considering in some detail

Loss of voluntary power.

Destruction of cortical substance or corticospinal pathways results in the failure of movement patterns being transmitted to the involved limbs. Recent work has increasingly emphasised the importance of cerebral movement patterns in the voluntary activities of the limbs. (Walshe 1923). When these patterns are destroyed voluntary movements of the limbs are grossly impaired; when they are damaged voluntary power in the limbs is reduced. As is seen in cases of mild hemiplegia it is the voluntary movements that the child achieves last that are most frequently and easily impaired. Thus

fine /

fine or rapid or independent finger movements or movements of the toes are always impaired in hemiplegia. The hand tends to be used all in one piece, all the fingers together, in an infantile fashion. Releasing the grasp is more difficult than grasping. In addition to impairment of finger movements it is usual to find impairment of the power of supination of the forearm and dorsiflexion of the feet at the ankle in even slight cases of hemiplegia.

In more severe cases there is more than weakness of movement, there is loss of the range of movement. Loss of the last few degrees of voluntary supination of a forearm was found to be an invaluable quick method of detecting mild cases of hemiplegia. The movements found to weakened in mild cases and limited in extent in those more severely affected were, in order of frequency, abduction and extension of the thumb, supination of the forearm, abduction and extension of the fingers, extension of the wrist and elbow, abduction of the shoulder, in the upper limb. In the lower limb voluntary movements of the toes were very much impaired in all but the mildest cases of hemiplegia then dorsiflexion of the ankle, extension of the knee, abduction and extension of the hip were less commonly affected.

Contracture.

In the early stages of active, acquired hemiplegia absence of some degrees of voluntary movement may be demonstrated /

Fixed contracture of the right foot in a position of equinus in a case of severe right hemiplegia with associated mental defect.

demonstrated in the limbs in most cases, though these movements may be full when the limb is handled passively. (Twitchell 1951).

Unless these movements are frequently actively attempted or accomplished passively a change in the state of the limbs occurs. Little used or neglected movements become impossible either actively or passively because of contracture, commonly appearing from 3 to 6 weeks after the appearance of hemiplegia in acquired cases. Because there is a tendency to neglect to utilise impaired voluntary movements to the full, restriction of movement due to contracture usually over-shadows the restriction of movement due to loss of voluntary power.

Contracture is therefore most likely to be extensive and severe in those with severe loss of voluntary power, who cannot move their limbs, in mentally defective or very young children who cannot be made to use their limbs or in those in whom no attempt is made to keep the limb mobile.

That the liability of contracture to affect little used limbs is not fully appreciated is indicated by the numbers of mentally defective patients encountered in institutions whose neurological disorder is hemiplegic but whose contractures are generalised. Because of the paralysis of one side the child finds movement difficult. Because he lacks desire to move he is allowed to reach the state where he cannot move because of contracture of the sound as well as the damaged side.

Case 48. Male. Born 1944. /

Case 48. Male. Born 1944.

Right hemiplegia with mental defect, aphasia and cortical blindness following a fit at the age of 10 weeks.

The first of two children born to mother aged 24 who suffers from epilepsy and father who shows and whose family shows syndactyly. Both highly intelligent.

The second child born at term after normal pregnancy and delivery is well.

During her first pregnancy with the patient the mother was well and the delivery was at term after a labour of 24 hours under chloroform analgesia.

The child appeared to be normal at birth, weighed 7 lbs. 10 ozs. and cried at once. The neonatal course was normal. From the age of 6 weeks he sucked the right thumb in preference to the left. He showed a slight right abducent weakness shortly after birth (evident in photographs). The only abnormality of possible significance was that when held up his legs crossed so that the grandmother wondered if they were abnormal. He was vaccinated at the age of 16 weeks, without apparent ill effect. At the age of 18½ weeks he was found head down in a soft pillow in his pram apparently dead. After some hesitation his medical grandfather applied artificial respiration and the child began to breathe after 15 minutes, during some of which time the heart sounds had been inaudible.

He seemed tired and drowsy for about 8 hours but otherwise normal. At the end of this time his face began to twitch on the right side and the twitching persisted, spreading to all limbs bilaterally, intermittently for 48 hours, after which the child again seemed rather drowsy but otherwise normal. He was slightly fevered.

The attacks of twitching occurred 2-3 times a day from this time onwards lasting a few minutes only on most occasions and involving the face and one or more limbs on different occasions.

He showed no attempt to hold the head up but was interested in what went on around him. He had begun to use the left thumb to suck instead of the right and the parents noted that he played with toys using the left hand only. He kicked with the right leg less than the right. At the age of 11 months, 6 months after his initial fit he had a severe bout of generalised convulsions lasting intermittently for 2 days and following them the parents noted a very definite deterioration in his vision which was progressive in the ensuing weeks.

Some /

Some progressive form of encephalitis was suspected and phenobarbitone prescribed. With the regular administration of phenobarbitone his fits ceased.

By the age of 2 he was able to sit without support and had said his first words. The right arm showed the position which it has retained, of flexion at the elbow, pronation at the forearm and flexion of the fingers. His right face moved less than the left. When he was induced to move the right hand spreading of the fingers and extension occurred which he did not seem to be able to control. He appeared to be able to appreciate light but his vision was limited and patchy.

He never acquired control of his bladder.

He was admitted to an institution for mentally defective children at the age of four. He has made little progress there. Is able to stand with support and say a few words, but he is unable to express more than his simple wants. Plays the mouth organ and enjoys hammering a piano. He is unable to use his limbs as freely as he was as owing to his perpetual sitting position he has developed contracture of his legs and slightly of the left arm as well as the right.

Father and his teachers have the impression that he understands rather more than he is capable of expressing.

Examination. Height 46". Head circumference 18". The head showed a very marked occipital flattening, in contrast to the slightly domed appearance of the frontal region. There was bilateral syndactyly of the 2nd. and 3rd. toes, slightly of the 3rd. and 4th. fingers. There was $1\frac{7}{8}$" shortening of the right arm compared to the left and 1" of the right leg compared to the left. He was easy to examine and helpful. He appeared to understand a fair amount of what was said to him. He was able to obey simple commands but his performance was inhibited by a great tendency to become frightened and upset. His speech is impaired and the consonants are slurred. His vocabulary is very scanty but is used appropriately. He is able to imitate quite complicated phrases, however, and occasionally he comes out with a jargon like word when attempting to reply to questions. Thus he repeatedly said "Hello, hello hello" when asked his name, though he said it perfectly well on later questioning.

The fundi showed bilateral optic atrophy, with large white discs. The blood vessels in both retinae seem to be few in number. He appeared to have some appreciation of light in the homonymous left upper quadrants and /

and to be able to distinguish large objects in that quadrant. There was a right abducent paresis. There was a right facial paresis of moderately severe degree and of upper motor neurone type.

There was a contracture of all the limbs. These are least in the left arm where pronation and extension of the elbow, only are slightly limited. They are most severe in the right arm, in which supination is impossible and extension of the elbow very severely limited. The right arm is not used independently but he uses both hands to handle objects, the right in an infantile fashion. The left leg shows flexion contraction of the hip, knee and ankle. The right leg shows more severe flexion contracture of the hip, knee and ankle. Movement is less, dorsiflexion of the ankle is very limited indeed and the contracture is more difficult to overcome. Superimposed on the contracture is a very marked spastic increase of tone in the right limbs with increase of biceps, triceps, supinator knee and ankle jerks, a positive right Hoffmann sign and extensor plantar response.

The left arm shows only rigid, without spastic, increase of tone. The left leg on the other hand does show some increase in the knee and ankle jerks and the plantar is probably extensor.

Sensory findings. Not testable in detail but light touch appears to be appreciated in all areas and no definite pass pointing or Rombergism could be elicited.

The right hand and foot are much colder than the left and show cyanosis. At times they are swollen.

On the other hand the degree to which function may be retained and contracture prevented by adequate physiotherapy and exercises is illustrated in the following case.

Case 176. Female. Born 1947.

Severe left hemiplegia. Congenital origin.

Only child of healthy parent. Mother aged 29 at time of delivery. Mother was well during pregnancy until the 8th. month when she developed some swelling of the ankles, the hands and face. She attended an antenatal clinic and mild pre-eclamptic toxaemia was diagnosed.

Labour commenced at 43 weeks and initially pains were /

were poor and irregular but after 24 hours pains became established, regular and good. The child was delivered spontaneously, vertex presentation, after 10 hours. The placenta showed very many infarcts. The child weighed 7 lbs. $3\frac{1}{2}$ ozs. and was limp and cyanosed. She responded to lobeline injected into the cord, and condition on remo al to the ward was stated to be satisfactory, and neonatal course, thereafter, normal.

The first abnormality noted was at the age of 2-3 weeks when the mother found that the left hand was difficult to wash because the fingers were clenched and the whole arm felt stiff. It was moved less than the right. Both legs appeared normal. Played with toys with the right hand only, from the age of 3-4 months. Sat without support at about 7 months and stood with support at about 1 year. Walking without support was considerably delayed, however, and she was 20 months before this was accomplished. When she walked she dragged the left leg and walked on the left toes. Talking was delayed until the child was aged 2 years but then developed rapidly and seemed normal.

She was seen by a clinic at the age of 11 months and considerable contracture of the arm and leg were noted to be present. From this age she received physiotherapy. Gradually the fingers were straightened one by one, and with encouragement she began to use the arm at the age of two, first to assist the right and then alone, in spite of great inco-ordination of the fingers. The position of the foot was improved by serial plasters and night boots and the gait improved parallel with the improvement in left hand manipulation.

She went to normal school at the age of 5 and is stated to be rather slow, but to be keeping her place.

Examination. Height $43\frac{1}{2}$". Head circumference 20". The left arm is $2\frac{1}{8}$" shorter than the right and the leg $\frac{7}{8}$" shorter than the right. There is a great reduction of the circumference of the calf and forearm on the left compared to the right.

Observant and probably of normal intelligence. Speech normal. Vocabulary good.

No facial asymmetry detected on rest or on movement.

The power of the left side is very much diminished in arm and to a lesser extent in the leg. Co-ordination was more impaired than power. Finger movements were very restricted and accompanied by great athetosis, spreading and extension of the fingers on the left. The toes /

toes cannot be moved voluntarily.

She is able to grasp with the right hand by utilising the athetotic spreading and extension for opening the hand and then allowing the natural following flexion of the fingers to occur.

Supination is almost absent as a voluntary movement and there is considerable pronation contracture. Elbow extension is very restricted. Considering the weakness of voluntary movements in the left upper limb, her use of it is surprisingly good.

The tone of the left limbs is much increased and spastic. The biceps, triceps, supinator, knee and ankle jerks are much increased on the left and the Hoffmann and Babinski responses are positive. There is left ankle clonus.

There are very marked flexor associated movements of the left arm when the right is in action or when she walks and relatively little response to tonic neck reflex.

Sensation. Slight diminution to discrimination between hot and cold and sharp and blunt in the left arm only. No disturbance of position sense and vibration sense is intact.

In cases showing severe contracture it is impossible to gauge to what degree apparent inability to move an affected limb is a true reflexion of the extent of the underlying loss of movement patterns.

The position of limbs showing contracture was very constant. The limbs took up positions of flexion because the extensor muscles tended to be more affected by paresis than the flexor muscles. The distal parts of the limbs were more affected than the proximal and more primitive movements were less affected by contracture than those of later development. Thus the thumb tended to be flexed and adducted into the palm, the fingers flexed, the wrist and elbow flexed, the forearm pronated and /

and the shoulder adducted and slightly flexed. The toes tended to be clawed, the foot to be held in a position of equinus and sometimes varus, the knee slightly flexed and the hip adducted and flexed.

Contractures limiting the voluntary movements of more complex type and later development were those most difficult to overcome. Adduction and flexion of the thumb, extension of the fingers, supination of the forearm, plantar flexion of the foot were the most resistant to treatment.

Cases with much involuntary movement tended to show less contracture than those in whom involuntary movement was slight or absent, but even the former rarely escaped some degree of contracture of the ankle, thumb, fingers and forearm.

Lower motor neurone lesions.

Lower motor neurone lesions associated with hemiplegia paresis have been largely ignored in recent years yet they were well recognised as being an occasional occurrence in hemiplegia in childhood by earlier authors. (Benedickt, 1874; Charcot, 1887; Brissaud 1880). With the increasing attention paid at the turn of the century to poliomyelitis, however, cases showing lower motor neurone lesions came to be classified as cases with mixed polioencephalitis and poliomylitis. (Lamy, 1894; Rothman, 1931; Clark, 1912). In the present series muscle reactions were performed on 6 cases of hemiplegia and /

and in four of these, chosen because lower motor neurone lesions were suspected the presence of these lesions was confirmed.

The cases were suspected because of the more marked wasting of the limbs than was usual in hemiplegia, because of the rigid rather than spastic increase of muscle tone and the marked dwarfing of the limb which was apparent . In two of the patients the lower motor neurone lesions were apparent in the muscles of the lower leg, in the peronei of one and the peronei and anterior tibial muscles of the other. In the other two cases the lesions were apparent, in one in the trapezius and deltoid and in the other in the sucinator and extensor muscles of the forearm. All the cases were classified as severe hemiplegias.

Case 24. Male. Born 1947.

Moderately severe left hemiplegia with lower motor lesion of peronei, following otitis media.

The first born of two children to healthy parents. Mother was aged 21 at the time of birth. The second child was born after normal pregnancy and delivery and is well.

She was well during pregnancy and the child was delivered spontaneously by the vertex in hospital after a labour of 11 hours, under chloroform analgesia.

The birth weight was 7 lbs. 1 oz. He was born in a state of blue asphyxia but responded rapidly to oxygen and thereafter appeared normal. The neonatal course was uneventful and he was discharged from hospital 7 days after birth.

Mother could not remember the early milestones but he /

he was crawling and sitting without support about the age of 9 months and was enjoying being bounced on his feet.

At the age of 9 months he began to rub his head on the pillow as if in pain and otitis media was diagnosed and treated with sulphonomides. Over a period of about a week he became more ill, more fevered, fretful, sleepless and anorexic. On the 10th. day after the onset of his illness he had a convulsion confined to one side of the body but the mother does not remember which. Following the fit the left leg and arm lay useless and were not moved. He was admitted to hospital for observation as a suspect case of poliomyelitis. In hospital acute bilateral otitis media was diagnosed and he was treated with penicillin for two weeks. When discharged at the end of this time he seemed "far away" , lazy and disinterested. The left side of his face sagged and the left arm was bent up and not used. He did not crawl or attempt to pull himself up as he had done before his illness.

He walked without support at the age of 15 months. When he did so he was unsteady, walked on the left toes and dragged the foot. The left arm was more bent up than at rest when he walked. He had babbled at the age of 9 months and words were spoken very shortly after his return home, without apparent retardation resulting from his illness. He had physiotherapy from the time of his discharge from hospital and his walking has steadily improved with splinting and calipers. He has been using the hand to assist the right from the age of $3\frac{1}{2}$. He went to normal school at the age of 5 and his progress has been better than average there.

Examination. Height $41\frac{1}{4}$". Head circumference 20". .3" shortening in the left arm compared to the right and $\frac{3}{4}$" shortening in the left leg. Considerable wasting of quadriceps, and peronei in the left leg with $\frac{1}{2}$" reduction in the circumference of the limb.

An intelligent responsive and interested child. Speech normal. Vocabulary good. There was a definite left facial paralysis of the upper motor neurone type on voluntary movement. On emotional movement on the other hand there was left facial overaction. At rest the face was symmetrical.

The power and co-ordination of the arm and leg was impaired on the left. The voluntary movements were full in range except for some limitation of supination of the forearm, extension of the elbow, but especially fine movements and extension of the fingers were weak on the left. Rapid movements of the left fingers were impossible.

He /

He was able to grasp and release objects, however. When he used the left hand spreading extension athetosis of the fingers is evident.

The tone of the left arm is much increased compared to the right. The biceps, triceps, supinator jerks are much increased. The Hoffmann sign is absent. The fingers are bilaterally "soft" and hyperextensible but this is more evident on the left than the right.

There was considerable generalised wasting in the left leg. This was especially severe, however, in the quadriceps and peronei. Lateral movement of the foot and toes was absent. Dorsiflexion was carried out entirely by the tibialis anterior and there is a tendency to varus deformity. The limbs showed an increase of tone which is a mixture of spasticity and rigidity. The knee and ankle jerks are much increased compared to the right and there is ankle clonus. The left plantar is extensor, flexor on the right.

The gait shows the effects of the spastic and the lower motor neurone lesions. There is a marked tendency to equino varus and the foot is dragged, toe down as he proceeds. When he walks or uses the right arm there is a tendency for the left arm to flex at the elbow, pronate at the forearm and for the fingers and wrist to flex. Extension of the elbow can be produced by use of the tonic neck reflex.

Sensory. No abnormality could be detected in superficial or deep sensation and no Rombergism or pass pointing was elicited.

On testing, the electrical reactions were shown to be of the lower motor neurone type in the left peronei and anterior tibial muscles.

It is hoped to perform electrical muscle reactions in a very much larger number of cases of hemiplegia in the near future. Unfortunately it is impossible at present to say how frequently lower motor neurone lesions occur in hemiplegic paralysis in childhood at present.

The importance of lower motor neurone lesions from the practical point of view is that they give rise to a form /

form of contracture whose successful treatment will not result in any improvement of voluntary power in the affected muscles.

According to early pathological work the origin of the lower motor neurone lesions lies in the spinal cord where small nodules of fibrosis spread from the degenerate lateral tracts to involve the cells of the anterior horns. (Brissaud 1880).

Dwarfism in hemiplegia.

In contrast to diplegic paralysis generalised dwarfing is not a feature of hemiplegia in childhood. On the other hand localised dwarfing of the limbs of the affected side has been noted from the time of the first studies of paralysis in childhood. (Morgagni, 1762 Cazauvielh 1827).

Interference with growth on the side affected with hemiplegia appears to have held a certain fascination for a number of German authors during the last century.

While French clinicians were adding to their knowledge of the effect of hemiplegia on the genitals and on fertility (Leblais, 1894; Gibotteau, 1889), large series of figures were collected and very accurate ideas obtained of the types and extent of dwarfing in hemiplegic German and American children. (Peterson and Fisher, 1889; Förster, 1880 ; Benedikt, 1874).

As /

As a result of these studies it soon became apparent that the extent and severity of dwarfism varied greatly in different cases. The trunk and face were involved in only a proportion of cases; some dwarfing of the limbs was usual but not invariable; the relative dwarfing in the arm and leg varied very greatly. At the same time little progress was made in discovering the reason for the dwarfing, though various suggestions were put forward to account for the phenomenon. One of the most plausible, based on the results of some of the investigations of Charcot and Bouchard was that dwarfing was in some way related to the degree of cord involvement (Brissaud 1880)

Dwarfing of the limbs.

In this series of 75 cases only 7 showed no apparent shortening of the limbs. Measurements were not obtained in 4 cases, in 3 because of overactivity of the children. The remaining 61 cases all showed dwarfing of the affected limbs to some degree. Measurements were taken in the arm from the tip of the acromium to the tip of the middle finger and in the lower limb from the anterior superior iliac spine to the internal malleolus. The degree of dwarfing found in the upper limbs of hemiplegic patients is shown in Table V.III.

It is possible to compare easily the degree of shortening only in a rather rough and ready manner owing to /

TABLE VIII

The apparent shortening in the affected arm, from acromion to top of middle finger in 75 cases of hemiplegia in childhood

Severity of the hemiplegia	Average age Yrs.	Mnths.	No. of cases	No shortening	Less than .5"	More than 1.5", less than 1".	More than 1" but less than 1.5"	More than 1.5" but less than 2".	2" and over	Unknown
Slight	8	9	27	7	6	8	3	1	0	2
Moderately Severe	7	9	26	0	6	9	7	3	0	1
Severe	9	6	22	0	2	4	4	7	4	1
Total	8	3	75	7	14	21	14	11	4	4

to the difficulties in allowing for the various differential factors of growth, at different ages and in the different sexes in the three groups of hemiplegic patients. Analysis in detail would be an excessively complicated matter. However, the differences in age and sex distribution between the slight, moderately severe and severe cases are relati ely small and it is clearly not possible to account for the much higher proportion of severely dwarfed limbs in the severely paretic group on this basis. Though it is apparent that, in general, the severity of dwarfing tends to be in proportion to thd severity of paresis, this is not invariably true. Thus, 4 cases of mild hemiplegia showed more than 1 inch of apparent shortening in the affected arm, and 6 cases of moderately severe paresis showed dwarfing of less than .5" compared to the unaffected arm.

Similarly though in general the severity of the dwarfing of the lower limb ran parallel to the severity of the dwarfing in the upper limb, this was also subject to variation. In most cases the apparent shortening from the anterior superior iliac spine to the internal malleolus in the affected compared to the unaffected leg was half that found in the upper limbs. Some cases showed almost equal shortening in the upper limbs and the lower limb and in others the difference between the sound and unsound upper and lower limbs was very marked. /

marked. In three cases of severe upper limb dwarfing the difference between the lengths of the upper limb was more than four times that found in the lower.

The reduction of the girths of limbs was usually proportional to the apparent reduction in length, even when lower motor neurone lesions were present. In some cases where tendon lengthenings had been performed or calipers had been applied the reduction of girth in the lower leg was relatively more severe than the apparent reduction in length.

In general it was difficult to assess to what degree dwarfing of an upper limb impaired function as severe dwarfing was usually associated with severe or moderately severe paresis, but in the leg it was of considerable importance. Though some compensation of reduction of lower limb length was achieved in many cases by the foot taking up an equinus position, walking was still abnormal when this was present. Not only does shortening of one lower limb result in an unsteady and clumsy gait but also to scoliosis of the spine and a thoroughly unnatural posture being assumed to counteract its effects.

What became increasingly apparent as the survey progressed was how much more acutely conscious of dwarfing of the limbs some patients were than of actual paresis. This was especially marked in adolescent girls whose attempts at camouflaging "The baby arm" were often elaborate /

elaborate and always pathetic. One remarked that the only thing she wished for was for her arm to become normal for one evening so that she could go dancing in a strapless evening dress. Such extreme sensitiveness about the abnormal limb obviously tended to make the patient use it to a less extent than was actually possible, and favoured the spread of contracture.

Vasomotor disturbances in the limbs in hemiplegia.

The temperature changes in children suffering from hemiplegia were found to vary greatly, as might have been expected from perusal of the older and the more recent literature. (Vulpian, 1875; Richards, 1947; Chevallier, 1867).

The bitter argument about the changes, so fierce in France in the latter half of last century is now less bitter but ideas on the subject appear to be but little clearer.

In this survey temperature changes were not measured except by the back of the hand in comparison to the normal limb. Therefore the conclusions are in the nature of impressions rather than the result of scientific observation.

In the acute stage of acquired hemiplegia it was frequent to find the affected limb warmer than the opposite side, the skin congested, the pulse fuller and sometimes oedema of the foot and hand was noted.

In /

In one case the congestion of the limb and the swelling of the hand and foot was sufficient to suggest a diagnosis of acute lymphoedemato to the attendant surgeon.

After a few days, in most cases the congestion of the limbs disappears, the oedema is absorbed and the limb becomes less warm. At the same time it begins to show excessive and somewhat erratic responses to changes in temperature, very readily becoming much warmer or much colder than the normal side. After a period of some weeks or months in the majority of cases the instability of temperature control is succeeded by a stage during which the limb is nearly always colder than the normal limb but in which it may be made very much warmer than the normal side by immersing some part of the body in warm water.

Some cases seem to recover from the initially vasomotor disturbance of their acute hemiplegia completely and temperature regulation seems to be grossly normal. A few cases show the permanently high temperature of the limbs noted so frequently in the acute stage however. Two such cases were observed during the series and both were symptomless. Examples of cases showing the exaggerated reaction to heat as well as cold were encountered quite frequently but were classified with those showing a reaction to cold as cold was the climatic condition which impaired the limbs function.

In /

In most cases these stages are ill defined and they merge into one another, but at the same time the alterations in the type of vasomotor reaction may easily be observed in individual cases. In the majority the end result is that the affected limb is colder than the normal limb in ordinary temperatures. It tends to become very much colder on exposure to temperatures below the usual, and may become cyanosed and swollen in even mildly cold weather. There may or may not be over reaction in the limb to warm conditions.

This is the most commonly encountered type of disturbance in the hemiplegic limbs and for purposes of classification they have been divided into three categories, mild, moderate and severe. Mild cases are those in which cyanosis is sometimes evident. Severe cases are those in which cyanosis tends to be persistent or very readily produced and in which oedema sometimes results from cold.

Case 142 is an example of the latter type.

Case 142. Female. born 1949.

Severe right hemiplegia with very severe trophic changes in the limbs, following convulsions at the age of 7 months.

The second of four children born to healthy mother aged 22 at the time of delivery and father who is also healthy. All family are right handed.

The third child was born at term by the breech.

All /

All other pregnancies and labours were normal and the children healthy.

In her pregnancy with the patient the mother was well. The child was born in hospital at term after an easy, normal labour of 1 hour. Delivery was spontaneous under chloroform analgesia. The child cried at once and seemed normal. Birth weight was 8 lbs. 8 ozs. The neonatal course was entirely normal.

Following her discharge from hospital the child seemed normal. She smiled at about one month, sat with support at $5\frac{1}{2}$ months and at the age of 6 months was playing with toys with both hands, recognising people and making noises.

At the age of 7 months she became out of sorts, refused food and refused to sit up. After 3 days the symptoms became more marked and she lay listless, taking little notice of her surroundings, irritable and was fevered. She remained in this state for 2 days and on the second of these the mother thought that her vision was impaired, and she seemed drowsy. She then had a convulsion which began suddenly with loss of consciousness and twitching of the face and the right arm which persisted for 10 minutes. She was adm tted to hospital where slight continuous twitching of the right arm only, without impairment of consciousness was noted. Papilloedema of the eyes was noted. Lumbar puncture findings were entirely normal. The following day it was noted that the right limbs were moved less than the left and she was very drowsy and fevered. Papilloedema persisted and she remained fevered. She was treated with penicillin and gradually over the course of a week the papilloedema settled and the temperature settled. At the same time the signs of a right hemiplegia became evident. This persisted during the six weeks during which she was in hospital. On discharge the mother found her extremely irritable and uncontrollable. She cried a great deal and would not be pacified. She was domineering and this behaviour persisted until the age of 2, since when it has gradually improved.

When she was discharged from hospital she "seemed to have been set back" so far as her development was concerned. She would sit up but for a long time made no effort to stand. She stood with support at the age of 18 months but was very unsteady and took her first steps without support only at the age of 22 months. She began to speak late also. She said her first words a few days after she took her first steps, at the age of 22 months. Her speech was poorly pronounced and the improvement in pronunciation has been slow. She added words very slowly and is still able to say only simple sentences.

When /

When she was discharged from hospital she did not use the right hand at all but with physiotherapy she began to use it to assist the left after 9 months treatment. After 18 months treatment she was able to grasp objects and at the age of 4 had learnt to pick up objects and release them with the right hand.

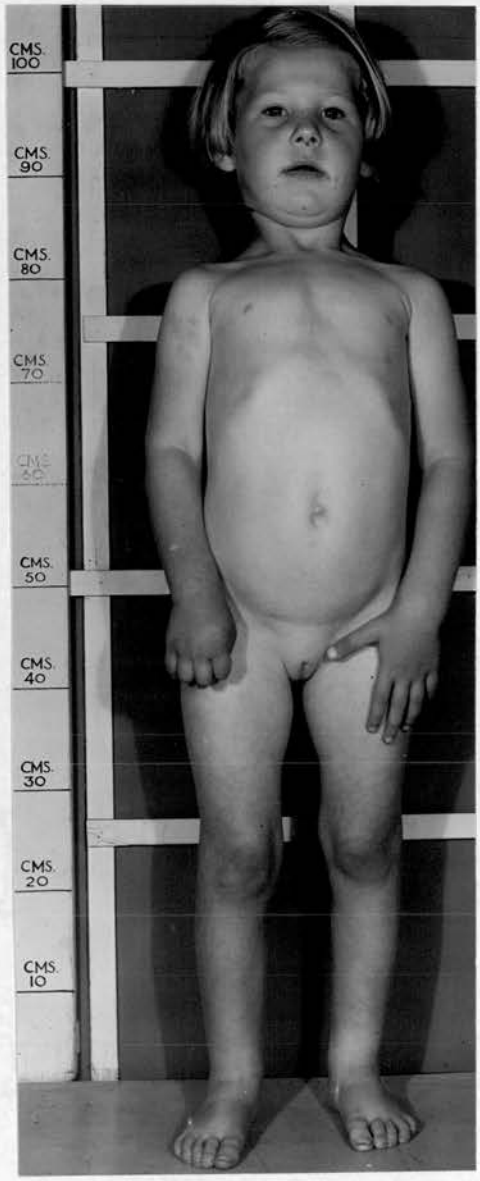
Though still unsteady her gait has steadily improved and she falls seldom. At the age of 3 she began to go to nursery school and is expected to go to a normal school at the age of 5, being regarded as normally intelligent.

During the second winter after her illness the right hand was noted to go blue and sometimes black in the cold weather. At times it was very swollen and she complained of pain in the hand. The right foot shows similar phenomena but in this the findings were less marked.

Examination. Height 40". H.C. $18\frac{5}{8}$". The left side of the head measured $\frac{3}{4}$" less than the right. The right arm was $1\frac{7}{8}$" shorter than the left and the right foot showed $\frac{7}{8}$" shortening compared to the left. She showed slight 2-3 syndactyly of the toes.

She was intelligent and replied to questions accurately, fully and well. Her response to commands was less good and she often did the wrong thing. Thus she persistently shut her eyes when asked to touch her nose with her finger, having been asked to do the former a few minutes before. She appeared to show a degree of aphasia though whether this was receptive or motor could not be determined. When she was asked questions requiring single word answers the answer was frequently out of place. Her speech showed poor vocabulary and marked hesitation. No stutter was present. Her pronunciation of all consonants was rather poor. Her response to imitation was extremely quick and accurate. There was a right facial paralysis of moderate severity and of upper motor neurone type. The power and co-ordination of the right arm were grossly reduced, that of the leg to a less extent.

Movements of the arm and hand on the right are very limited. There is only a few degrees of supination of the forearm, the fingers cannot be fully extended and the thumb cannot be fully lifted from the palm. Extension of the wrist is limited to below 150' and extension of the elbow and abduction of the shoulder is weak. Nevertheless she is able to pick up objects, grasp them and release them. All movements of the limbs are accompanied by quite marked athetoid movements of the fingers /



Case I40. Marked dwarfing and vasomotor changes in the upper limb in a case of acquired right hemiplegia. The cyanosis and oedema of the distal parts of the arm will be noted.

fingers with classical extension and abduction of the digits, associated with extension of the wrist. She utilises this movement to increase the range of finger extension when she wishes to grasp an object and the voluntary extension of the fingers is inadequate.

There is limitation of dorsiflexion of the right foot and she is unable to move the right toes voluntarily. Abduction at the hip and extension of the knee are weak.

When she walks she dips to the right and tends to walk on the right toes, dragging the foot as she proceeds. The right arm is strongly contracted, adducted at the shoulder, flexed at the elbow, wrist and fingers and pronated in the forearm. The right arm also takes up this position when the left arm is used.

The tone of the right limbs is spastic. The tone of the right arm is much increased, that of the leg moderately. The biceps, triceps, supinator, knee and ankle jerks are much increased on the right compared to the left and the right Hoffman sign is present. The right plantar is extensor, the left flexor. The right hand shows great hyperextensibility and softness of the fingers.

Sensory. Absolutely no deficit of the cutaneous sensation or of joint or vibration sense could be discovered on the right side, though to simultaneous pin prick bilaterally that on the right is ignored.

The right limbs were much colder than the left and the hand and foot were slightly cyanosed and 'blotchy'.

When the opposite hand was placed in cold water the right hand and foot could be observed to become almost black in colour, swollen and so stiff as to be almost incapable of movement.

Of the 75 cases of hemiplegia in the series, 16 showed no gross disturbance of temperature regulation in the affected hand. 39 cases showed slight disturbance, 10 showed moderately severe disturbance and 6 showed severe disturbance. Two cases showed an increase of temperature in the affected arm, compared to the normal which /

TABLE IX

Vaso-motor changes in the limbs of 75 children with hemiplegia.

Severity of hemiplegia	No. of cases	Affected limb colder. Degree of difference			Affected limb Always warmer	No difference in limbs	Unknown
		Slight	Moderate	Marked			
Slight	27	12	2	0	0	13	0
Moderately Severe	26	16	3	1	1	3	1
Severe	22	10	5	5	1	1	0

which was permanent and apparently no handicap.

Of the cases classed as being severe all except one, were also classed as cases of severe paralysis; of those classed as suffering from moderately severe vaso-motor disturbance two were classed as mild cases of paresis, 3 as suffering from moderately severe paresis and 5 as suffering from severe paresis. It will be observed that the figures in Table IX. indicate that, though in general the incidence and severity of vasomotor disorder bears some relationship to the severity of the paresis, this is by no means always the case. One case of severe paresis showed no apparent difference in the temperature of the limbs and in another the affected hand was permanently warmer than the other.

The importance of the vasomotor disturbance in the limbs in children with hemiplegia appears to be somewhat underestimated. In moderately severe and severe cases the affected limbs may readily become so cold that their function is severely impaired during cold weather. The contrast in the performance of a child in a slightly cold room with one in a normal or slightly warm one is striking. It was no accident that one child, made to use the affected hand at school, remarked that he got good marks for writing in summer but bad marks in the winter.

In some of the more severe cases pain as well as loss of function results from the exaggerated response to cold /

cold of the damaged limb.

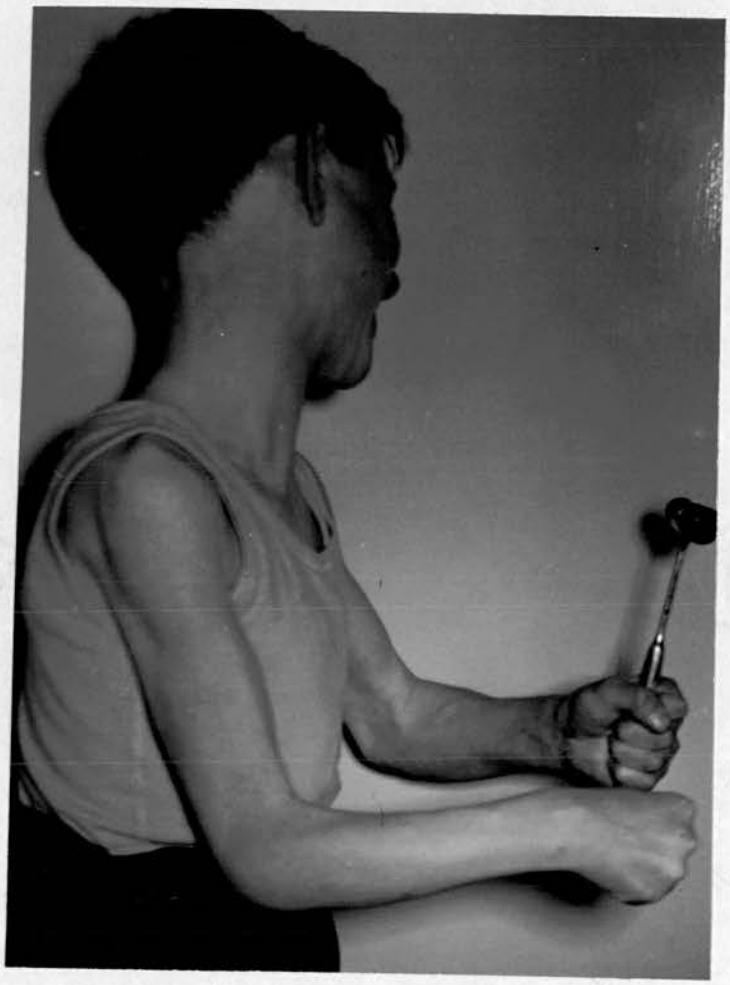
Involuntary movements in hemiplegia.

In addition to limitation of function of the affected limbs due to paresis function is also altered by the presence of abnormal movements in affected side. These are generally described as being of three types and are all of very frequent occurrence in hemiplegia in childhood.

The first type consist of generalised associated movements, "syncinesie globale" which are in the nature of uninhibited postural reactions occurring in the hemiplegic limbs as a result of exertion of any type. In general they tend to "produce the typical attitude of predilection (contracture)", in the affected side. (Mondrad-Krohn, 1948). These movements are almost always present in cases of hemiplegia.

The second type are imitative movements in the affected limbs found when the contralateral limbs are in action, "Syncinesies d'imitation". These movements are rather less common than those of the preceding type.

The third type of movements are those resulting from defective muscular co-ordination in the affected limb itself, resulting in disordered movement when a voluntary attempt is made to move it. These are commonly called "Co-ordinated associated movements", of "syncinesies de co-ordination", neither of which terms /



Case 167.The associated flexion movements in a case of moderately severe right hemiplegia.The patient is grasping the hammer forcefully in the normal left hand.Marked flexion movement of the right upper limb is apparent.

terms is particularly adequate as a description of the variety of disordered voluntary movements encountered.

Generalised associated movements.

The presence of these movements has been recognised for many years and a number of early authors described them accurately and fully. (Todd, 1854; Cazauvielh, 1827; Cruvehlier, 1862; Raymond, 1876; Charcot, 1887; Audry, 1888). That the movements were the manifestations of primitive postural patterns normally inhibited in those with intact cortico spinal systems, but uninhibited when this system was damaged became apparent though the neurophysiological work done at the turn of the century. (Sherrington, 1905; Magnus and de Klejn, 1926).

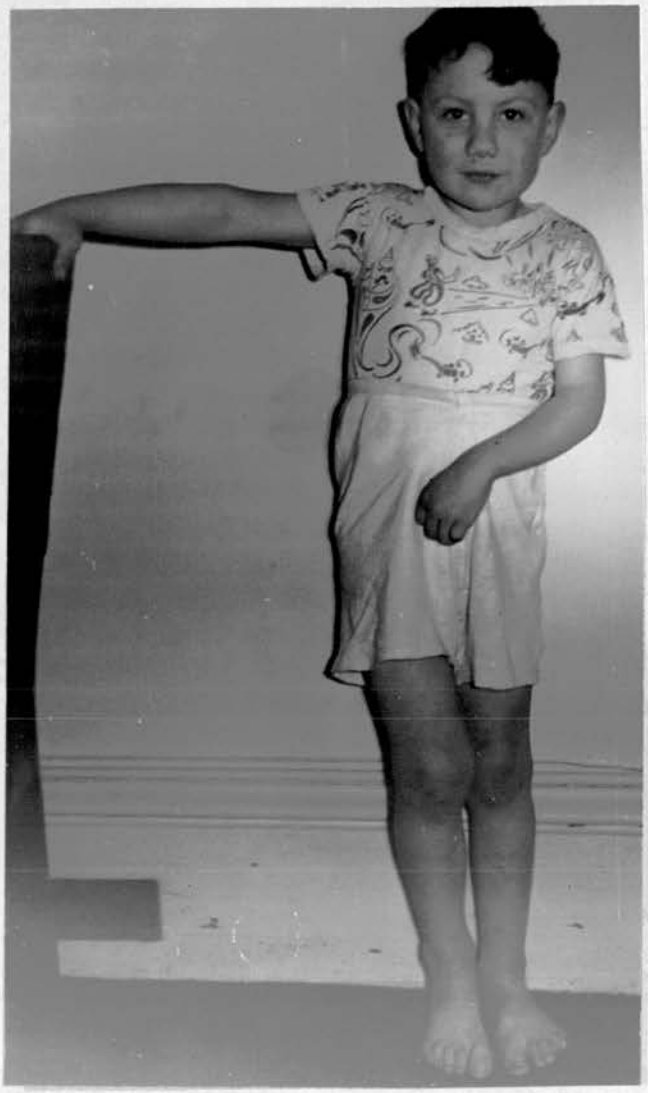
That they are in the nature of righting reflexes, easily demonstrated in decorticate animals has been shown, and the importance of the position of the head in relation to that of the body in altering the position of the affected limbs has been emphasised.

In this series involuntary movements of the generalised associated type were apparent in every case of hemiplegia. The severity of the movements and the ease with which they were elicited depended to some extent on the severity of the paresis in the limbs. If the child exerted himself sufficiently, especially with the head turned to the hemiplegic side, so that the tonic neck reflex came into action, they could always be produced, even /

even in the youngest and mildest of cases of hemiplegia. The existence of these movements was someti es of diagnostic help.

The activities which tended to produce the generalised associated movements were those in which the child exerted hi self to any considerable extent. In severe cases of hemiplegia they might be produced by acts as slight swallowing or by the child becoming excited. In slight cases of hemiplegia more strenuous activity might be required, running, lifting a weight with the unaffected arm with the head towards the hemiplegic side, or by means of performing the tonic neck reflex briskly. In most cases the associated movements were more easily produced with the child standing than lying, with the head extended than flexed, and after he had had some exercise of the affected limbs.

The movements of the affected upper limbs were predominantly flexor in type all but two of the cases of hemiplegia examined fully. When the child exerted himself he showed adduction at the shoulder, flexion at the elbow, wrist and finger, pronation of the forearm and adduction of the thumb. The findings in the lower limbs varied more in most cases flexor positions were assumed, contrary to the usual statements of the findings in adults; the hip adducted and the hip and knee flexed. The /



Case 93. Associated flexion movements of the affected upper limb in a patient with left hemiplegia. The boy is grasping forcefully with the right hand.

The foot took up a position of plantar equinus so that the shortening of the limb resulting from hip and knee flexion was to some extent compensated. In 27 of the 74 cases examined in detail, extensor positions were assumed in the leg on exertion, sometimes with hyper-extension at the knee. In most cases, therefore, exertion tended to precipitate an exaggeration of the position in which the limb was held as a result of contracture. The tonic neck reflex in hemiplegic patients affords another clue as to the nature of the reflexes which result in the associated movements. In the vast majority of patients it was positive, but the movement it produced in the limbs varied very greatly. In those with marked contracture when the head was turned to the normal side, the tendence to extension of the affected arm was slight, but in most cases a very marked extension took place. In mild cases and those in whom contracture had been prevented the arm could frequently be made to take up a position of adduction and internal rotation at the shoulder, extension at the elbow, pronation of the forearm and flexion of the wrist and fingers. This decerebrate like position of the arm was associated with a marked increase of tone in both affected limbs and always with a degree of extension in the affected leg, frequently with slight hip flexion.

The /

The degree to which the limbs responded to the tonic neck reflex was dependent upon the severity of the contracture present in the limbs, but also upon whether the positions of associated movement in the limbs tended to be strongly flexor or not. Thus in the cases where running with the head held straight produced markedly flexor patterns in the paretic arm the tonic neck reflex usually produced only slight extension when the head was turned to the sound side. In the two cases in which normal exertion produced only extensor positions in the affected arms very little flexion was produced by turning the head towards the affected side.

The importance of the generalised associated movements from the point of iew of the effects of their function is two-fold. In the first place in slight cases of hemiplegia acts requiring the use of two hands or of the paralysed hand when the child is walking or otherwise exerting himself, are made difficult by the tendency for the affected limb to assume the position of prediliction, usually the position of flexion. It is very noticeable that children taught shoemaking are more successful in schools where they do not have to work the sewing machine with their legs, than where they do.

The generalised associated movements are important in the second place because to some extent the child may be /

be able to utilise them. Thus one boy was able to hold a piece of wood for sawing in a barrel making factory, by means of putting the wood in the affected hand with the unaffected and then grasping his saw with much more exertion than was really required to hold it. The exertion was sufficient to flex the fingers of his damaged arm and hold the wood, the flexion in the elbow of the affected arm was utilised to wedge it against the sawing block.

Imitative movements.

Descriptions of imitative movements in the literature are very frequently movements of the generalised associated type, postural mechanisms whose true nature has not been recognised. True imitative movements are not nearly so common and were observed to some degree in only 15 cases in this series, 9 of them slight cases of hemiplegia and 4 moderately severe. The movements most commonly imitated are unfortunately those tending to place the limb in the position of prediliction, usually flexor positions. Imitative movements are more likely to occur in movements involving considerable exertion on the sound side. The simplest test is to ask the child rapidly to pronate and supinate the sound arm and watch the affected limb. It will usually be found that the generalised associated movement predominates and that a position of steady increased flexion

is /

is achieved but occasionally the wrist of the affected limb may be seen to extend and flex slightly. The affected limb also appears to be more likely to imitate movements arising as a result of emotion. Thus when the sound arm is suddenly raised in alarm to ward off a blow, the affected arm may follow suit. Unfortunately, since voluntary movements of extension and supination are much less frequently imitated, (though they are occasionally) imitative movements are of little functional value to the child.

The following patient showed the rather unusual picture of some imitation of extensor positions.

Case 134. Male. born 1938.

Severe left hem plegia. Arrested hydrocephalus.

The 12th. of 13 children born to healthy parents. Mother aged 41 at time of delivery. 4th. child died in convulsion at the age of 3. The 10th. and 11th. were twins, second was a breech presentation and died shortly after birth. All remaining children are healthy and mostly in trades or apprentices. Youngest bursary winner.

All weights were between 7 and 8 lbs. at birth except for twins and last born, who was $9\frac{1}{4}$ lbs. All except twins spontaneous deliveries at term after normal labours.

In her 11th. pregnancy with the patient she was well. Delivery was at term after normal labour of 8 hours. It was spontaneous. Child cried at once and appeared to be normal. Weight was $8\frac{1}{2}$ lbs. Normal neonatal course.

First abnormality noted was at about 4 months when it was seen that his left arm was held by the left side "in a useless kind of way". At 5 months a health visitor pointed out to the mother that the child's head was growing very rapidly and was already much bigger than it should be.

He /

He sat up unsupported at the age of 9 months and walked without support at the age of 16 months. Had some words by the ti e he walked. When he began to walk it was noted that the left arm was held tightly to the side and bent at the elbow with the hand clasped tightly over the thumb which was always in the palm of hand. His gait was "lop sided" and he dragged the left leg and wore out the toe of his left shoe. Following a fall at the age of two he had a period of vomiting and headache for 3 weeks but this passed off. Went to school for physically handicapped at the age of 5 and did moderately well there. I.Q. 85.

With operative lengthening of tendon achilles on 4 occasions his gait has improved somewhat but he still limps and his leg has become steadily thinner. Can use the left hand for holding objects but cannot release grasp or use it for much independent activity.

Examination. Height 67½". H.C. 23¼". Well built adolescent. 2" shortening in the left arm and 1" shortening in the left leg compared to the right.

Slow in response to command or to questioning but quite accurate. Has little imagination but some sense of humour. Speech shows good vocabulary but his pronunciation of all consonants is poor and his speech is jerky. No stutter.

Left facial paresis of upper motor neurone type. The power of the left arm and leg is reduced, the former to a greater extent than the latter. Co-ordination is more impaired than power. Movements of the left arm are very limited. There is practically no supination and elbow extension is very limited. He has no power of finger extension voluntarily. With any movement of the left hand there is very marked extension athetosis of the fingers. He utilises this involuntary extension to grasp objects, but is unable to release them. There are also imitative associated movements and by extending the wright wrist and fingers some extension can be produced in the left. Similarly by holding the right elbow straight some extension may be produced in the left.

There is very great increase of tone of a spastic type with some contracture rigidity in the left arm compared to the right and a similar, less marked increase in the left leg. The left fingers are hyperextensible. The Hoffmann signs are negative. The left plantar is extensor, the right flexor.

His gait is defective. He dips to the left and drags the left toe, still does not get the heel to the ground. When he walks the left arm is markedly flexed and the forearm /

forearm pronated. By use of the tonic neck reflex, however, the arm can be made to take up a position of extension at the elbow as far as the contracture will allow and forearm pronation.

Sensation. Entirely normal sensation on detailed testing by hot and cold test tubes, sharp and blunt and vibration senses. No proprioceptive loss demonstrable. The left hand and arm are somewhat colder than the right.

Imitative movements are generally relatively weak and very readily limited by contracture.

Athetosis and other forms of co-ordinated associated movements.

Inco-ordinate movements of the fingers in some cases of hemiplegia were noted by early workers interested in hemiplegia on the continent and in this country. (Todd, 1854; Cruvehlier, 1862). Following the detailed description of these movements and their being named athetosis much interest in involuntary movements in hemiplegia was aroused. (Hammond, 1881). Numbers of descriptions of movements of different types appeared, athetoid, choriec and mixed in type. These were discussed and related in a masterly paper by Gowers (1874). More recently there has been a tendency to stress the occurrence of abnormal movements and to ignore the paresis, which is the more severe cause of disability, both in hemiplegic and in diplegic forms of cerebral palsy.

In this series athetosis was found to be the rule rather than the exception in cases of hemiplegia. By athetosis is understood a condition in which voluntary movements of the affected limb, especially of the distal /

distal parts, accompanied by slow, inco-ordinate writhing movements of the fingers, and less commonly the wrist and the forearm. Similar phenomena may be observed in the toes and the feet in most cases.

Athetosis was of a rather similar pattern in most cases though the extent and severity to which the limbs was involved varied greatly in different cases. It was most commonly produced by movements of the affected arm which required a mixture of postural activity and voluntary muscular activity. The clumsiness of the affected hand in most cases of hemiplegia means that its activities are confined to grasping objects. Delicate manipulation is impossible.

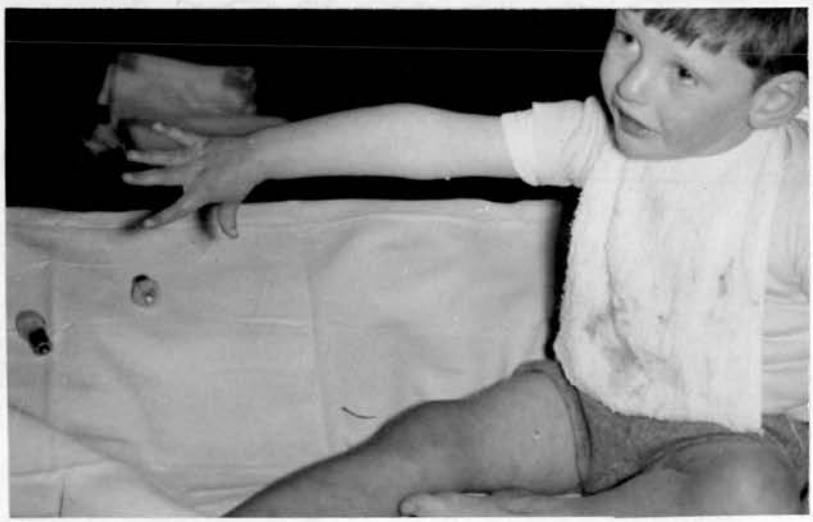
TABLE IXa

Severe athetosis in hemiplegia in childhood.

Severity of hemiplegia	No. of cases	No. showing severe athetosis
Mild	27	4
Moderately severe	26	5
Severe	22	10

It is most common when the child reaches for objects, and is maintaining the limb in a steady position prior to grasping the object with the hands, that athetosis /

Case I29.Severe athetosis of the affected upper limb in a patient with severe left hemiplegia.



Case I84.A patient with right hemiplegia.Severe athetosis evident at the moment of voluntary grasping with the affected hand.

hemiplegia were those which showed athetosis of the limbs of this degree. But a smaller proportion of slight cases did also.

Though it would be a handicap, as it was in the majority of cases who showed it, athetosis could also be utilised by the child to increase the use of the affected hand. The cases in which athetosis occurred most frequently were those in which paresis was severe enough to have lim ted the power, accuracy and extent of voluntary finger extension more or less severely. Children so affected found that grasping objects was difficult largely because they could not easily extend the fingers enough to grasp objects. Releasing objects was usually equally difficult.

By means of utilising the full extension of the fingers found in athetosis a number of children, quite severely handicapped by their hemiplegia were able to open the hand enough to grasp objects and release them with surprising facility.

Case 190. Male. Born 1938

Moderately severe right hemiplegia with marked athetosis.

The first of two sons born to healthy right handed parents. Mother was aged 22 at the time of delivery. Paternal uncle and aunt and two cousins are left handed.

Second child born after normal pregnancy and labour is well.

During her first pregnancy with the patient the mother was well except for some vomiting in the early months. The delivery was at term in hospital and was spontaneous, after a labour of 13 hours. The presentation was vertex and the delivery accomplished under chloroform /

chloroform analgesia.

The child cried at once and seemed normal. Weight was 6 lbs. 6½ ozs.

Apart from some initial difficulty in fixing to the breast the child seemed normal. At the age of 3 months the mother noted that he was using the left hand but not the right and that the right was difficult to wash because the hand had to be opened before the palm could be dealt with. The hand seemed to become stiffer rather than less stiff during the ensuing weeks and right hemiplegia was diagnosed at the age of 4 months.

The child held the head up at the age of 2 months, sat with support at the age of 6 months and without support very shortly afterwards. He never crawled but stood without support at the age of 13 months and walked without support at 14 months.

When he began to walk the right toes were inturned and he did not put the heel to the ground. The position of the arm had tended to be flexed across the chest before the age of 1 year but walking seemed to exaggerate this position. He resented physiotherapy to attempt to correct the position of the arm and gradually the limbs became more persistently fixed in the position of flexion though with constant encouragement he did make some efforts to move it from the age of 4. His gait showed little improvement and the tendon achilles was lengthened operatively at the age of 4. Thereafter the gait improved but again deteriorated after the age of 7.

His speech was rather slow in developing. He said his first words at the age of 18 months but added to them slowly. His pronunciation was slow and laboured but he never stuttered. He went to a normal school at the age of 5 but was transferred to a school for the physically handicapped at the age of 8.

He was slow to learn to read and write. He was better at mental than written arithmetic. He reversed letters and numbers persistently until over 12 years old and if tired or in a hurry is still liable to do so. He has always tended to invert sentences, use words out of place and occasionally seems to absolutely stick, being unable to comprehend quite simple commands. He lost the sight of his right eye by running into a dart at the age of 4. He had a very severe septicaemia at the /

the age of 7, during which he was desperately ill.

Examination. Height 67⅕". H.C. 22⅕". 1¼" shortening of the right arm and ¾" shortening in the right leg compared to the left. Adolescent.

Slow in response to questions or commands. Willing but sluggish. Expressing himself is much slower, however, than one would expect from the degree of his comprehension. He shows some word replacement traits which he failed to notice himself in ordinary conversation. No abnormality of pronunciation noted.

Amaurotic right eye, which shows abducent palsy.

There was a right facial paresis of moderate severity and upper motor neurone type. The power and co-ordination of the right arm were poor. Voluntary extension of the elbow, very weak and limited. Shoulder abduction was weak. Rapid and fine finger movements were quite impossible. He was able to grasp and release objects by utilising the very marked extension athetosis of the fingers which w as present. This was evident on the least attempt to move the limbs and involved also overextension of the right wrist. When he misjudged the athetosis objects were liable, even when he had succeeded in grasping them, to fly from his hand as if thrown. In general the right arm was only of use as a ste dying agent for the left. Movements of the leg on the right were much less impaired, though some passive and voluntary limitation of dorsiflexion of the foot, extension of the knee and abduction of the hip was evident. His gait was abnormal, the knee and hip held partially flexed and the foot in a varus position and plantar flexed. When ever he used the left arm or walked a marked degree of adduction at the shoulder, flexion at the elbow, wrist and fingers, pronation of the forearm was apparent. The tone of the right arm and leg was spastic and markedly increased. The biceps, triceps, supinator, knee and ankle jerks were all markedly increased and the right plantar was extensor, the left flexor. The right Hoffmann sign was positive, the left negative. The tendon jerks on the left were normal, and co-ordination and power very good.

Sensory. Superficial sensation was normal over the right limbs. There was some loss of accurate proprioception with the right hand and foot but vibration sense was normal. The right hand and foot were much colder than the left, though no cyanosis was apparent.

Choreoid movements./

Choreoid movements.

Choreoid movements, were much less frequently encountered than athetoid in hemiplegia in childhood, but a large number of cases in the earlier literature show that their occurrence cannot be considered rare. Cases of choreoid movements affecting the hemiplegic limbs were reported by (Raymond, 1876; Charcot, 1887; Bouchaud, 1894; Audry, 1892; Gowers, 1874). Various transitional forms of disorder occur, in which involuntary movements of athetoid as well as choreoid type may be found.

In type the choreoid movements that occur are interesting in that essentially they consist of a rapid intermittent series of involuntary movements which tend to bring the limb into one of the positions of predi-liction. Usually the position eventually assumed is one of extension of the upper arm rather than flexion but the position of the limbs fluctuates widely and markedly between the positions of flexion and extension when voluntary movement is attempted.

Case 143. Male. born 1940.

Moderately severe right hemiplegia with hemichorea and athetosis. Congenital origin.

First and only child born to mother aged 42 and healthy father. Mother had much nausea and vomiting in early months of pregnancy and persistent backache later on. Labour began 6 days after expected date. After 36 hours there was little advance and a comiciliary attempt at forceps extraction failed. Thereafter admitted to hospital where considerable oedema and elevated blood pressure were noted. Morphia was given and a very difficult mid-cavity forceps delivery accomplished /

accomplished under anaesthesia. The placenta was offensive. The child's birth weight was 10 lbs. 8½ ozs. The child was limp and cyanosed at birth but responded slowly to resuscitation. Respiration irregular for ½ hour. Thereafter loud cerebral cries. Right facial palsy noted, and both right limbs noted to be weak. Child remained irritable, restless and slightly cyanosed for three days and then began to improve, the remainder of the neonatal course being normal.

All milestones normal except that walking without support was delayed until the child was 2 years old and was then noted to be clumsy and thought to be ataxic. Child had always been left handed and only began to use the right hand to assist the left at the age of 6 and alone at the age of 8.

Speech started before the age of 1 and has always been slightly slurred, with the consonants poorly pronounced.

Examination. Height 59½". H.C. 21.4". 1½" shortening in the right arm and ½" in the leg compared to the left. Head elongate but not asymmetrical.

Rather slow in thought and speech but accurate and original. Speech - poorly pronounced consonants.

Astygmatic and myopic. Slight right facial palsy of the upper motor neurone type evident only as lag on emotional or voluntary movement.

The power of the right arm and leg is much reduced. The elbow cannot be extended voluntarily beyond 45'. Supination of the forearm and dorsiflexion of the ankle actively and passively severely limited. Fine finger movements are impossible. The toes cannot be moved voluntarily.

Whenever he makes any voluntary movements, associated movements of the right arm and face take place. They are maximal in the region of the right shoulder. The movements are quick and jerky in type and result in a throwing of the head to one side, the arm from, and then to the right side. The movements start with broad swinging, choreoid like jerking of the arm which is followed by a succession of smaller jerks which appear to spread down the arm and to the face as a twitch, extending from the face to the fingers. As the movements decrease the tone of the right arm increases and the limb assumed a flexed position at the elbow, pronated at the forearm, flexed at the wrist and fingers and adducted at the shoulder, the hemiplegic position. As he walks these movements are persistently apparent though more marked when he attempts to use the arms, especially the right. When he does attempt to move the right arm there is athetosis of the fingers ... a spreading movement with over-extension and then flexion of the fingers, as the limbs, /

limbs, again, assumed the "Hemiplegic position".

The tone of the right side is much increased and the jerks are accentuated but there is no clonus. The Hoffmann and the Babinski are positive on the right.

Sensory findings. A lack of hot and cold and sharp and blunt discrimination, and some impairment of joint sensation is apparent on the right side. The right hand and foot are somewhat colder on the right than the left.

Even slight choreid movements tend to handicap the function of the affected limb considerably because not only are the fine distal movements of the limbs affected, but the movements of the proximal joints, those designed to maintain the position of the limb steady, also tend to be impaired.

As in the other forms of dyskinesis to be discussed later choreoid movements complicating hemiplegia tend to be exacerbated by stress, excitement and infections.

Neglect of the paralysed side.

Neglect of the paralysed side is a phenomenon which appears in the acute stage of hemiplegia and persists to a variable degree when the acute stage disappears. Since it occurs following epileptic fits in some cases neglect of the paralysed side has been regarded as being in the nature of a cortical exhaustion phenomenon. Since it occurs in a number of cases with sensory impairment of the limbs it has been regarded as being a result of damage to cortical sensory areas.

In fact the explanation for the very apparent neglect of the limbs, also observed in chimpanzees with precise sectioning of their pyramidal tracts, remains wanting. /

In general all the patients with acquired hemiplegia in whom a full history was obtained, a history of neglect of the affected arm in the acute stage was elicited. This persisted to a variable degree afterwards in different cases. The neglect tended to be more severe the more severe the paresis of the limb, but not uniformly so.

Because neglect of the limb leads to its disuse and favours the onset and spread of contracture it is an important cause of permanent loss of function. It is also a disability that can be largely overcome by making the child use the limb consciously as much as possible. A good mother will be found saying almost reflexly whenever the child does anything, "Now use the right hand", "No with the other hand", many hundreds of times a day. She may have to continue with this for many years.

Sensory findings in those with marked limb neglect were not dissimilar to those in whom neglect was not evident. In both marked sensory abnormalities were absent. At the same time when the affected and unaffected limbs were pricked or otherwise similarly stimulated, simultaneously the affected limb was almost always ignored, though on separate testing the pin prick would be said to be felt as sharply in both limbs.

Sensory impairment. /

Sensory impairment.

The classical authors found that sensory involvement occurred only occasionally in the involved limbs in hemiplegia. (Freud and Rie, 1891; Osler, 1889; Lovett, 1888; Charcot, 1887).

In this series some degree of loss of position sense was found to have occurred in almost all severe and a large proportion of moderately severe cases. The loss of position sense was found consistently to be more severe in those with immobile limbs than in those in whom movement was less impaired. The loss of position sense when it occurred alone, was slight in most cases. It was similar to the slight loss which was found in a number of patients whose limbs had been immobilised for long periods because of orthopoedic conditions or poliomyelitis.

It was possible to compare the findings in 11 children with approximately similar degrees of severe hemiplegic paralysis. 6 of these had received no, or merely routine inadequate physiotherapy. 5 had been treated as in or out patients in a school for spastic children where physiotherapy, and therefore active and passive movements of the limbs had been greatly encouraged. One of the five cases so treated showed superficial and deep sensory impairment in the affected limbs, but in the other four no cases were found with any /

any loss of position sense. In the 6 patients with relatively immobile limbs loss of position sense was demonstrated in all, though in every case it was only of slight degree.

Impairment of sensation other than the slight impairment of position sense alone, above referred to, were apparent in 4 cases. The majority of cases showing these changes were severe or moderately severe cases of hemiplegia.

Superficial sensation was impaired on testing in 10 cases. In most the impairment was slight. In 2 it was associated with joint sense of a greater degree than could be expected merely from the relative immobility of the limb. The cases showing impairment of superficial sensibility were all cases of moderately severe or severe hemiplegia.

Cerebellar ataxia was shown by 4 patients with hemiplegia. 3 of these cases were classified as being severe or moderately severe cases of paresis. 1 was classified as mild. The ataxia was manifested as pass pointing, more marked in the affected limbs and by deviation when walking forwards or backwards blindfolded. The ataxia was usually to the affected side, 3 of the 4 cases. It occurred more markedly towards the unaffected side in one case. It is probable that a mild degree of ataxia occurs in many cases of hemiplegia. It is almost always found that when the child stands with the feet together he is very much more easily pushed to one side than to the other. The side to which he is more

easily pushed is the paralysed one in only about half the cases.

In cases of hemiplegia in childhood with sensory impairment it was a matter of the greatest difficulty to estimate to what degree function of the limbs was impaired as a result of the loss of sensibility, since so man of the cases were suffering from severe or moderately severe vasomotor disability.

Other findings in hemiplegia.

Under this heading will be considered the other abnormalities encountered during the examination of the 75 cases of hemiplegia in the survey. Some of these are important from the point of view of impairing the patient's performance and limiting his capabilities in a variety of ways. Other findings were of more clinical interest than practical importance.

Intellectual impairment.

The assessment of intellectual ability in cases of cerebral palsy is a matter of the greatest difficulty. In the first place the child may be incapable of showing his intelligence because of his motor impairment. In the second place his intelligence may not have had a chance to develop fully because of the restricted experiences and limited environment he has lived in. Not only may his environment and experiences be limited by /

by his motor handicap but the senses may be directly affected; some have specific difficulties in the comprehension and expression of their ideas, of aphasic type. Obviously such handicaps cramp the development of intellectual abilities.

On intelligence testing, therefore, performance may not be a true measure of the child's actual intelligence, because of his motor handicap. Even when it is a true measure of his actual intelligence it is no estimate of the intelligence which he may be capable of developing in an ideal environment. Thus intelligence tests can only be regarded as very rough guides to actual or potential intellectual capacity.

A patient's social and educational achievements are also poor measures of his intellectual capacity in cerebral palsy, for many severely paralysed cases with normal intelligence show marked retardation in their developmental milestones and in their educational and social progress.

Yet, although they are fallacious, the only useful measures of the child's intellectual ability that are available are the levels of his performance on intelligence testing and in social and educational achievement. Experienced observers are able to assess intellectual capacity /

capacity on a basis of these findings, in a large proportion of cases, though a long period of observation under standard conditions may be necessary. (Dunsdon, 1952).

In Table X. are shown the intelligence quotient found in a number of the cases of hemiplegia by various psychologists under varying conditions. In addition the levels of their educational achievement, as measured very roughly by the necessity to send them to schools for the physically or mentally handicapped or to institutions are shown. It will be observed that while a greater proportion of se erely paretic patients are of low intelligence than of slightly paretic patients, there is a gre t variation in the relative severity of the paresis and the intellectual impairment. The range of ariation extends from grossly mentally defective children with mild hemiplegia to cases of very severe hemiplegia with above average intelligence. That differences of kind, as well as of degree exist between these poles of the clinical spectrum is very likely. Unfortunately types of intellectual impairment cannot be expressed in neurological or neuropathological terms.

Examples of the extreme types of cases met with follow.

Case 117. Female. born 1938.

Mild right hemiplegia, mental defect. Convulsions in infancy.

Born /

TABLE X

The intelligence and schooling of 75 cases of hemiplegia in childhood.

Intelligence Quotient.	115+	100-114	85-99	70-84	55-69	Less than 55	Untested
Severity of the paresis							
Mild	1	2	4	4	8	5	3
Mod. severe	2	4	2	4	7	4	3
Severe	0	4	4	9	2	2	1
Normal schools	1	3	9	8	2	0	0
Schools for physically handicapped	0	1	2	4	3	0	1
Schools for mentally handicapped	0	0	0	6	8	1	0
Ineducable	0	0	0	0	1	6	2
Not yet at school	0	2	1	2	1	4	7

Born to a mother aged 32 and father aged 38 who had been married 8 years. Mother had a subsequent miscarriage. Patient is the only child. Both parents right handed and healthy. Mother was well in pregnancy and the delivery was at home, at term after a normal labour of less than one hour. The child was born spontaneously and no anaesthetic or instruments were necessary. The birth weight was 7 lbs. and the child seemed normal. The child seemed normal for the first 2 weeks of life and then vomiting with constipation began and persisted until she was five weeks old, when she was admitted to hospital. At this time the weight was only $4\frac{1}{2}$ lbs. she was grossly dehydrated and it was thought she might die.

From the age of 3 weeks she had had attacks of loss of consciousness, in which she became pale, went stiff suddenly and after about one minute appeared to be normal. These attacks occurred 5-6 times a day.

In hospital she received intravenous and subcutaneous drips for one week and then a Rammstedt operation for hypertrophic pyloric stenosis was performed. The recovery from the operation was uneventful and she began to gain weight. On discharge at the age of 7 weeks the weight was 5 lbs. The fits persisted, however, though they gradually became less frequent over the years, occurring once or twice a week when she was two years old and ceasing by the age of 4. They remained similar in character to the initial attacks.

When she began to handle objects the mother noted that the child used only the left hand and that the right seemed stiff when she washed it. On examination in hospital there was some limitation of supination of the right forearm as early as 4 months. She looked about her at the age of 4 months, held up the head about the age of 6 months and was sitting without support at the age of 1 year. She began to walk without support at the age of 15 months but she was very unsteady as she proceeded and the right foot was dragged. Until the age of 4 the gait remained unsteady but has improved since then. She made her first attempt at words shortly after the age of 15 months but her progress was very slow. The content of her speech was very poor and the pronunciation so bad that she was incomprehensible until the age of 3. Since that time the mother has understood a fair amount of what she says but her speech remains very backwards.

Since the age of 6 she has begun to use the right arm a little to assist the left, but still refuses to use it independently.

As /

As she has grown older she has appeared to fall further behind in intellectual development. She proved ineducable at normal school and since the age of 7 has attended an occupation centre. I.Q. said to be less than 50.

Examination. Height 59½". H.C. 18½". ½" smaller left hemicircumference than right. Gross occipital flattening. The right arm is 1" smaller than the left and the right leg shows ½" shortening. Well built adolescent. Very backward. Able to speak but only words, not sentences. Pronunciation is very bad and there is gross slurring and lack of differentiation of consonants which makes understanding difficult. She can count to five and say her own name but can do little more in response to questions.

There is a slight right abducent paresis.

The right arm and leg show impairment of power and co-ordination on the right compared to the left. This is most marked in the right hand where finger extension is very weak and clums . Supination of the forearm and extension of the elbow are full passively and there is no contracture but they are somewhat limited on voluntary movement. When she is made to move the right hand, especially for grasping, the fingers show extension and abduction of the fingers of a slow writhing and involuntary nature ... true athetosis. The tone of the right limbs is increased and is spastic in type. The biceps, triceps, supinator, knee and ankle jerks are increased compared to the left. The right Hoffmann is positive and the right plantar is extensor.

Shen she uses the left arm or walks the right arm assumes a position of flexion at the elbow, wrist and fingers and pronation of the forearm with adduction at the shoulder, taking up the flexed hemiplegic position.

The gait is clumsy, but not obviously spastic and there is no apparent limp though closer inspection shows that the right heel is hardly placed to the ground.

Sensory findings untestable. The right hand and foot are colder than the left.

Case 171. Female. born 1943.

Severe right hemiplegia, associated athetosis, Jacksonian epilepsy and sensory impairment. Congenital origin.

The /

The second of three living children born to Rh-ve mother aged 24 at the time, and father Rh positive. Both parents healthy.

First child born after a labour of 10 hours, at term, weight 8 lbs. Healthy. Third child, given replacement transfusion immediately after spontaneous delivery, on account of rhesus incompatability. Well. Two subsequent stillbirths with hydrops.

Pregnancy normal. No increase in antibodies found on routine testing of mother's serum. Admitted to hospital in labour and delivered spontaneously after 8 hours of patient, who weighed 8 lbs. 13 ozs. and was born face to pubis. Appeared to be normal at birth, and for 5 days. Then became jaundiced and seemed lethargic. Haemoglobin fell to 50% on 7th. day and blood transfusions, two via the saggitatal sinus were necessary in the next 10 days. During one transfusion strabismus, air hunger and a shrill cry were noted, and for 2 days following this transfusion there was persistent vomiting. Child was discharged aged 6 weeks and appeared to be normal. No note as to splenomagaly.

Child appeared normal on return home but the right leg seemed stiff and the child was always left handed, not using the right hand at all until the age of 3 or 4. Held head up at the age of 4 months, crawled at the age of 2 and walked without support shortly after the age of two. When she began to walk it was noted that the right leg was dragged and she walked on the toes. The right arm was held bent across the chest and used very little.

Following treatment at the age of 4 with splinting to arms and legs and serial plasters to the legs the walking improved and the use of the right arm increased. The child's squint persistent from infancy has improved steadily.

From the age of 3 she had fits. These begin with a cry, great impairment, but not loss of consciousness and rigidity of the right hand, arm and leg. There is no great jerking during the fit and she may or may not be incontinent during them. She frequently complains of tingling in the right arm and leg immediately before or immediately after the fit. They last 10 minutes usually, but occasionally longer and have only been partially suppressed with phenobarbitone.

She goes to normal school and does quite well there. She had some difficulty in writing letters and figures from the age of 6½ to 8½ years, but read well /

well and has no difficulty in writing now.

Examination. Height 53". H.C. 19½". There is 1¾" shortening in the right arm compared to the left and ¾" shortening in the right leg.

A nervous, tense child, normally intelligent. Alert and co-operative. Speech entirely normal and of good content. I.Q. 112.

Severe abducent paresis of the lefteye. Slight droop of right angle of mouth at rest, but movements normal. Power of the right limbs is much impaired. The impairment is severe in the hand and co-ordination is very greatly impaired. Finger movements are very slight and accompanied by extension athetosis with spreading on the least attempt at voluntary control. There is considerable contracture of the arm inhibiting voluntary elbow extension, forearm supination and wrist extension. Dorsiflexion of the right ankle, full extension of the knee and abduction of the hip are also limited.

With any voluntary movement of the left arm or when the child walks, there is a marked tendency for the right arm to take up a flexed and typical"Hemiplegic" position. On the other hand the position of extension of the elbow, adduction of the shoulder, pronation of the forearm and flexion at the wrist and fingers, "the decerebrate position" is very readily obtained by the tonic neck reflex or by forceful activity of either leg when she lies down.

The tone of the right arm and leg is spastic and increased. The biceps, triceps, supinator, knee and ankle jerks are much increased and the Babinski and Hoffmann responses are positive.

Sensory. There is some apparent increase of sensitivity on the right side to pin prick and hot and cold, more marked in the arm than the leg, compared to the left. Other modalities, joint position and vibration sense appear normal. The right hand and foot are much colder than the left and the hand and foot are frequently, though not persistently blue.

That the success attained in education is not entirely dependent either on intellectual capacity, or on /

on the severity of the paresis is illustrated by the wide range of intelligence quotients and severity of of paralysis found in children in similar types of schools and institutions.

In cases of hemiplegia of congenital origin the milestones are usually retarded, but the actual milestones which are retarded and the degree of retardation encountered varys greatly. In those who have much mental impairment, but little paresis the milestones are usually all late, sitting, walking, talking and manipulating. In those in whom there is severe paresis but little mental defect it is usually found that sitting is somewhat retarded, walking more or less severely retarded but that the child beg an to handle with the unaffected arm and to speak at about the normal time, even if pronunciation is defective. But the variations encountered in the behaviour of cases similar in the degree of their mental impairment and their paresis is great. Moreover cases are encountered in whom the intellectual defect appears to improve during the years, and others in which it appears to become worse. Similarly in acquired hemiplegia the degree of intellectual impairment apparent in the acute stage gives very little indication of what the eventual intelligence will be. In some cases the intelligence appeared to improve for three or four years after the initial illness.

Case 191. /

Case 191. Male. Born 1946.

Slight right hemiplegia following measles encephalitis.

The only child of healthy parent, both right handed. Mother was 20 at the time of delivery. She was well in pregnancy and was delivered in hospital at term after a labour of 7 hours, spontaneously under chloroform analgesia. The baby was very large, weighing 9 lbs. 14 ozs., but appeared to be normal and cried immediately after delivery.

He appeared to be normal in infancy. He sat with support at the age of 5½ months and was using both hands to handle toys before this. He stood without support between 8½ and 9 months and walked shortly afterwards. His speech was rather slow, he said his first words at the age of 18 months but then it developed very rapidly.

He was rather a quite boy.

At the age of 5 he developed measles and 6 days after the development of his rash, when he had seemed to be improving he became very drowsy, lost his appetite and complained of headache. The doctor found he had neck retraction and that he was very fevered. He was admitted to hospital where a diagnosis of measles encephalitis was made. He appeared to be confused, unable to obey commands and not fully oriented for the first week of his stay. The following week he was drowsy and vacant and had one slight generalised convulsion with loss of consciousness. Lumbar puncture findings were normal. Thereafter he improved and was discharged after 9 weeks in hospital.

For about 1 week in hospital he seemed to be unable to see objects so that he could recognise them, unable to understand speech according to the parents.

On discharge his whole nature was changed. He was demanding and could brook no frustration. His temper was labile and variable. He was overactive and curious, never still for a moment. His speech was ill pronounced and most of it impossible to understand because he ran words into one another. He made no effort to use the right hand which was "held cramped up against the chest". He limped and dragged the right foot.

In the year following his discharge from hospital his overactivity became less and his speech improved so /

so that only when he is excited is it now incomprehensible and his walking became steadier. After 6 months he began to use the right arm to assist the left. After 18 months he was able to do buttons with the left hand assisted by the right. He attended normal school after 6 months at home and has done quite well there being in the middle of his class with children of his own age.

Examination. Height 45½". H.C. 21/. The right arm was ⅝" shorter than the left. The legs were of equal length.

He was alert and quick in response to commands or questions but not always accurate. He tended to be facile rather than clever. He showed flights of ideas related only by chance word associations. His speech was normal. His comprehension was good.

There was a right facial paresis of upper motor neurone type, moderate in degree. The power and co-ordination of the right arm and leg were impaired. The grasp was moderately good and large objects were firmly held. On the other hand, small, fine or rapid finger movements were very poorly performed. All movements of the right hand were accompanied by marked athetosis of the ring and middle fingers which extended and separated. The athetosis was much less in the index and little fingers and was slight in the wrist, so that the athetosis did not grossly impair his efficiency by itself.

The only movement limited in extent on voluntary effort was supination of the forearm but abduction of the shoulder, tension of the elbow, wrist and fingers were weaker than on the left. In the leg dorsiflexion of the foot and knee extension were weaker than on the left.

The gait was abnormal with the foot being in a position of slight equinus and the knee slightly flexed. When he walked or used the left arm the right tended to assume a position of adduction at theshoulder, flexion at the elbow, wrist and fingers and pronation of the forearm.

There was moderate increase of tone of spastic type in the right limbs and the biceps, triceps, supinator, knee and ankle jerks were increased on the right compared to the left. The right Hoffman response was present, the left absent. The right plantar response was extensor, the left was flexor.

Sensory findings were similar and normal on the two sides.

The right hand and foot were considerably colder than the left.

It is not proposed to discuss the tremendous

diversity /

diversity of the findings in cases with impaired intelligence. The findings of almost enc clopoedic memory in some, could be compared to apparent absence of memory in others, the good social adaptation of some with negligible educational achievement and the complete social dependence of some cases with relatively little intellectual impairment. The variations encountered were so numerous, and the factors involved so complicated no short review of them is possible.

Apart from in itself being a tremendous handicap, intellectual impairment also had the indirect effect in the majority of cases showing it, that it led to the paretic limbs being used to less than the extent to which they were theoretically capable, and because disuse was common the incidence of severe contracture was higher in the mentally defective than in those with normal intelligence.

Aphasia.

Interference with the reception and expression of ideas in speech was found relatively commonly in cases of hemiplegia. In those with mental defect of moderate or severe degree it was almost invariable, but overshadowed by the intellectual impairment. Cases in which the interference with the reception or expression of ideas was clinically recognisable, either because it was /

TABLE XI

Aphasia and speech difficulties in 75 cases of hemiplegia in childhood.

Hemiplegia	No. of cases	Aphasia	Speech defects			
			Dyslalia	Stutter and hesitation	No comprehensible speech	All types of speech defects
Right	54	11	8	3	2	13
Left	21	1	6	1	1	8
Slight	27	2	7	2	2	11
Moderately severe	26	6	6	1	1	8
Severe	22	4	1	1	0	2
Normal schools	23	1	0	0	0	0
Special schools	26	4	10	2	0	12
Ineducable	9	3	1	0	2	3
Under 5	17	4	3	2	1	6

was a relatively isolated or severe disability were classified as aphasic. There were 12 such cases.

In view of the controversy, especially rampant in the late years of last century as to whether aphasia was at all dependent on the side of the brain damage, it is interesting to note that all but one of the cases of aphasia showed right hemiplegia. The exception was left handed before the onset of his acquired hemiplegia. (Bernhardt, 1885; Marie, 1888; Dreyfous,1882).

Though the numbers are too small for any statistical treatment it is interesting to note that 8 of the 12 cases occurred in case of acquired hemiplegia. Many cases from whose histories it seemed likely that aphasia had been present in the acute stages of the acquired hemiplegia showed no such abnormality at the time of examination.

In some cases the degree of aphasia was severe and ability to imitate speech in these children was very striking compared to their complete inability to express their ideas. In milder cases the aphasia was more of a nuisance to the child because it resulted in frequent word misplacements and substitutions than a very serious disability.

In Table XI are shown the numbers of cases of aphasia which occurred in patients showing mild, moderately severe and severe paresis. It will be noted that aphasia occurs relatively /

relatively less frequently in cases of mild hemiplegia.

Case 192 showed severe aphasic speech disorder.

Case 192. Male. born 1948.

Severe right hemiplegia. Aphasia. Congenital origin.

The first of two children born to mother aged 28 at the time of delivery and healthy father. Mother's father is left handed. Mother had recurrent fits in childhood.

Younger child was born spontaneously at 43 weeks, after a labour of 48 hours. Well. She was well in her first pregnancy. Labour began with poor pains, 5 weeks after the expected date, at home. They persisted in all for 5 days, but only in the last 24 hours were the pains severe. "The baby got stuck" and had to be turned by the doctor and delivered with extreme difficulty by forceps. He showed marks from the forceps after birth and was limp and blue. He did not cry for over an hour. The birth weight was $8\frac{3}{4}$ lbs. Immediately after delivery the mother noted that the child's right hand w as clenched. He did not move the right arm or leg though he moved the left. He was irritable and difficult to feed. Gradually over the weeks he seemed less irritable but the right arm became bent and stiffer as time went on.

At the age of 10 months he sat without support, and walked without support at the age of $2\frac{1}{2}$. He was unsteady, limped and dragged the right leg as he proceeded. He spoke normally.

Taken to hospital at the age of 10 months because he would not use the right arm and physiotherapy and splinting was prescribed. He began to use the right arm to help the left at the age of 3 and occasionally for holding things at the age of 5.

Went to physically handicapped school where his progress has been slow.

Examination. Height $41\frac{1}{2}$". H.C. $19\frac{1}{2}$". Left hemicircumference $\frac{1}{2}$" less than right. Syndactyly of 2-3 toes bilaterally. The right arm is $1\frac{1}{2}$" less than the left and the leg $\frac{3}{4}$" less in length than the left. There is 1/3" difference in the hemithorax, the right being smaller. Child plays intelligently with toys. When excited the well constructed sentences, of which he is capable, turn into single repeated words to his obvious distress. When speaking normally, however, there is still a tendency for curious phrase inversions and word replacements.

Whereas /

Whereas his obedience to command is usually brisk and accurate, his ability to name objects and to respond to some commands appears to "Block" sometimes, so that he stands gaping, unable to start to speak.

Right pupil larger than left. Vision normal.

Right third nerve weakness. Right facial weakness evident on voluntary movement. Tendency for head to be tilted to the right. Very severe paralysis of the right side. Thumb and finger movements practically absent.

Severe pronator and elbow flexor contracture. Abduction of the shoulder very weak. Has learnt to use the extensor athetosis of his fingers to grasp objects but cannot release. Leg less affected than arm but no movement of right toes and only slight, voluntary, dorsiflexion at the ankle.

Associated movements of the right arm are severe and consistently of the flexor type whenever the left arm moves or he moves the legs.

Both hands are "soft",the fingers are grossly hyperextensible.

The tone of the right limbs is greatly increased. The increase is largely spastic but there is an element of rigidity presumably due to the contracture. The biceps, triceps supinator knee and ankle jerks are increased moderately. The Hoffmann and Babinski responses are positive.

Sensory. Very difficult to test but no gross abnormality discovered.

Speech defects.

To a person trained to listen to speech very high proportions of hemiplegic patients are found to have defective speech, (Dunsdon, 1952). The proportion depend very much on the standards adopted as to what is normal and what abnormal speech. It is clearly necessary that some criterion of what is defect and what is idiosyncrasy should be stated. In this series minor abnormalities of pronunciation, slight enough not to interfere with the comprehension of the child's speech by normal people were ignored.

For the purposes of the survey only those whose speech was so severely defective that comprehension of it was made a matter of difficulty were included. Cases whose vocabulary was poor but pronunciation was normal, and who were able to make their meaning clear were not, therefore, included. Three cases of hemiplegia, all severe in type and two of them right handed had no speech or only a few ill pronounced words.

Fourteen cases suffered from multiple dyslalia of such degree that their speech was difficult to comprehend and four cases had stutters. Speech disturbances were found to be somewhat commoner in slight than severe cases of hemiplegia. Dunsdon's observations on all speech defects, tended to show that speech defects were more than twice as common in right hemiplegia as in left hemiplegia. She noted the slightest defects, however, and all that can be stated as a result of the present observations is that this was not true of severe speech defects, in the present series.

It will be noted that speech defects tended to be commoner in mild than in severe hemiplegia. This may be related only to the greater tendency for children with mental defect to show speech disorder, but at the same time, as has already been noted, there is a tendency in mentally defective patients for the hemiplegia to be less clearly defined. Cranial nerve involvement, ataxia, and some increase of the tone of the leg on the opposite side /

side to the hemiplegic occur more frequently in defective patients, suggesting that nervous pathways are in some ways more diffusely affected, in these children.

Case 15. Female. Born 1944.

Slight left hemiplegia. Mental defect following illness at 10 months of age.

The fourth of five children, all girls born when mother was aged 37. Mother died 18 months later with carcinoma of the uterus. Mother and two of the children showed syndactyly of the 2nd. and 3rd. toes. Youngest girl has thrombocytopenic purpura. All the children were born after normal pregnancy and delivery.

Mother was well in her fourth pregnancy. Delivery was spontaneous and normal after a labour of a few hours. Weight was 8 lbs. 7 ozs. She cried at once and seemed normal. Normal in infancy until the age of 10 months. By this time she was sitting alone and beginning to crawl. She was babbling and parents were listening for the first word.

Following a fall from her pram the father thought that the child's legs were abnormal, were used less and felt "rubbery" when moved. At about the same time he noticed a slight squint and that the face was twisted. She was admitted to hospital and discharged after one week, diagnosed as suffering from the effects of concussion.

She stood without support at the age of 14 months and began to walk shortly after. Her walking was very unsteady however; she was always banging into things and falling. She fell more to the left than the right and moved the left leg round in a circle when walking though she moved the right normally. After the age of two the walking began to improve and since the age of 5 she has fallen seldom and the gait appears "clumsy still but pretty normal."

In infancy she had used the left hand more than the right but following her accident she changed gradually over the course of 2 years and is now quite strongly right handed. She has always been able to use the left hand independently, however, though less neatly than the right.

Her speech was slow to develop, starting only at the age of 18-20 months and when she did speak the pronunciation was slurred. She also tended to run words together /

together and misplaced and substituted words, especially when excited. She went straight to a school for the mentally handicapped at the age of 5 and received speech therapy. Her pronunciation has steadily improved there but she still omits and misplaces words when she is excited. Her progress at school has been rather poor. She had persistent letter and small word reversals until the age of 8 and still occasionally muddles b and d. I.Q. 52.

Examination. Height 49". There is 1/3" shortening of the left arm and $\frac{1}{4}$" shortening in the left leg compared to the right. No asymmetry apparent in the head or trunk.

Response to simple commands good, but to complicated commands very poor. Speech shows some slurring of consonants but it was entirely comprehensible and no aphasia was noted.

Right eyed and right handed.

Slight right abducent weakness. Face shows a slight upper motor neurone lesion. The power of the hands is fair, somewhat impaired on the left compared to the right. The legs show power which is within normal limits. Fine movements of the fingers are much less good on the left than the right and extension of the fingers is weaker on the left. Rapid movements are similarly impaired. There is no restriction of voluntary movement in the limbs. These are full. She is able to use the left hand independently and to do buttons with the hand but handling is much clumsier than with the right though there is no apparent athetosis or other specific abnormality to account for the difference. There is a moderate increase of tone in the left arm and leg compared to the right. The increase is spastic in type and the biceps, triceps, supinator, knee and ankle jerks are all considerably increased. The Hoffmann signs are absent. The right plantar is flexor, the left is extensor.

Shen she uses the right arm there are associated movements in the left. The elbow flexes, the forearm pronates and the fingers clench. These movements are also seen in the arm when the child runs. Her gait is clumsy but not markedly abnormal. The toe cap of the left shoe is more worn than that of the right.

Sensory findings are entirely normal. The left hand and foot are colder than the right.

In /

TABLE XIa.

Amaurosis in 75 cases of hemiplegia in childhood.

Type of paresis	No. of cases	Complete amaurosis	Unilateral amaurosis	Field defect	Totals
Right hemiplegia	54	1	1	5	7
Left hemiplegia	21	1	2	1	4
Severity					
Slight	27	1	0	0	1
Moderately severe	26	0	1	3	4
Severe	22	1	2	3	6
Totals	75	2	3 x	6	11

x Two cases of unilateral optic atrophy are of postnatal injury.

In Table XI. are shown the numbers of cases with speech defect attending normal and special schools and considered ineducable. It is interesting to note that no child attending normal school had speech severely enough affected to make it difficult to comprehend. This may partly be due to the fact that such children, whatever their intelligence, tend to be transferred to special schools on account of their defective speech.

Of the cases showing stutter, one case of congenital hemiplegia developed it only at the age of 6 years. The other three cases showed stutter from the time they began to speak, or shortly after.

Visual Defects.

Visual defects of various types are common in hemiplegia, and were well recognised by earlier German authors and discussed in much detail by them. (Koenig, 1895; Freud, 1889).

The common defects in this series were of two types, errors of refraction and amaurotic. Little account was taken of the former, and unfortunately the exact incidence of refractive cases than in those of normal intelligence. Five cases of severe myopia were recorded. Four of these cases were mentally defective.

Of much more clinical importance are the amaurotic disorders. The types and numbers of these encountered are shown in Table XIa. It will be seen that severe and moderately severe cases of hemiplegia show amaurotic defects more frequently than do slight cases. Two cases of hemiplegia showed

no /

no useful vision. Both these cases were of congenital origin and in both bilateral optic atrophy had occurred.

Two of the cases showing unilateral amaurosis were of great interest. Both were amaurotic in the opposite eye to the side of the paresis. In both the retinae showed extremely scanty vessels and very white optic discs with irregular margins. The appearance was of severe unilateral optic atrophy probably the result of retinal thrombosis. Possibly the involvement of the retinal vessels was related to involvement of the cerebral vessels which could be held responsible for the hemiplegia .... a speculation too difficult to resist.

The cases showing field defects did so consistently to the side of the paresis. The degree of field defect varied greatly in extent. In one case epileptic fits commenced with phenomena appearing in the defective quadrant.

Case 5. Female. Born 1938.

Moderately severe left hemiplegia with epilepsy and hemianopia dating from birth.

She was the third of 4 children, born to mother aged 40 and her healthy husband. The mother had had her first child prematurely, delivered by forceps. The second child was born dead. The fourth was a normal delivery and the first and fourth are well.

She was well in her pregnancy with the patient until the week before delivery when she had a small vaginal haemorrhage. Delivery was spontaneous at home after a normal labour of 2 hours. It was approximately 3 weeks premature. The child cired at once and initially she seemed normal.

On her second and third days of life the child showed generalised attacks of twitching with loss of consciousness intermittently. /

intermittently. Thereafter the neonatal course was normal.

The child held up her head at the age of 4 months, sat with support at the age of 9 months, stood and walked with support at the age of 19 months. She said her first words shortly after the age of 2. When she began to walk the gait was noted to be clumsy and she walked on the left toes and dragged the leg.

At the age of 7 months it was noted that the left arm was stiff and moved less than the right. Later it became flexed at the elbow and was bent across the chest rather rightly whenever she walked.

She received physiotherapy from the age of 4 years and gradually her gait improved, the foot being dragged less and the heel being put to the ground. After the age of 6 she began to use the left arm to assist the right and since the age of 10, independently, though to a small extent.

At the age of 2 she began to have epileptic attacks once every 2-3 months. The attacks persisted in spite of phenobarbitone administration. During them she feels faint, sees bright lights, scenes or people, always on her left side, but does not lose consciousness. After 2-3 minutes the visual phenomena disappear and she feels well again. Sometimes during attacks she feels nauseated and occasionally vomits. They occur gradually enough for her to sit or lie down before faintness develops.

At the age of 5 she went to a school for the physically handicapped. She kept her place in class there but was rather clumsy. I.Q. 95.

From the age of 2 or three she has been noted to laugh and cry easily, "without really meaning it", says the mother.

Examination. Height 61". H.C. 19¾". There was ½" shortening in the left arm and ¼" shortening in the left leg compared to the right.

She was a well developed adolexcent girl. She showed a marked tendency to laugh and cry unemotionally, or at any rate, not in accordance with her mood.

Her speech was of good content but pronunciation was poor with slurred consonants but was comprehensible except when she was excited.

Left lower quadrantic homonymous hemianopia. Left pupil was larger than the right. Left third nerve paresis.

The /

The face was symmetrical at rest but showed a lag on the left side on voluntary movement and an overaction on emotional action. The tongue showed involuntary undulating movement on protrusion.

All movements of the limbs were clumsy, but there was much impairment of power and co-ordination on the left compared to the right. There was limitation of passive and voluntary supination of the left forearm, extension of the elbow, wrist and fingers. All movements were much weaker and find and rapid movements of the fingers impossible. She was able to use the hand to pick up objects and in order to achieve full finger extension, she utilises the marked athetosis of the fingers constantly evident on the least movement of the left arm. With the athetoid extension and separation of the wrist was associated extension of the wrist.

Her gait showed dipping to the left with the knee and hip slightly flexed on that side and an equinus position of the foot evident. When she walked or used the left arm, adduction of the shoulder, flexion at the elbow and wrist and fingers was evident. The forearm was pronated. With the tonic neck reflex the elbow extended.

The tone of the left arm and leg was much increased compared to the right and aspastic in type with moderate increase of biceps, triceps, supinator, knee and ankle jerks, compared to the right, though no colunus. The left Hoffmann sign was present and the left plantar response was extensor, the right flexor.

Sensory findings were normal. The left foot and hand were colder than the right.

Of the cases showing visual field defects, 4 were acquired in postnatal life. Of these four cases, three were right hemiplegics and showed aphasia in addition to their field defect.

Case 17. Male. Born 143.

Severe right hemiplegia, mental defect. Gross overactivity, focal epilepsy following cerebral abscess.

The first born of three children to healthy mother aged 22 and father of 36. Mother's father was left handed but rest of family right handed.

One miscarriage year before patient was born. Two last born children well after normal pregnances and deliveries.

During /

During her first pregnancy she had German measles at 4 months gestation. Thereafter she was well. The delivery was at term in hospital after a normal labour of 5 hours. It was spontaneous and the child cried at once and seemed normal. Birth weight was 7 lbs. 15 ozs.

The neonatal period was normal. The mother developed a staph. aureus breast abscess one month after the birth and 10 days later, aged 5 weeks, the child began to ail, lose weight, have severe diarrhoea and became dehydrated. Pneumonia was diagnosed and sulphonomide and saline infusions given with improvement. After 3 days, however, the child, whose temperature had failed to settle completely, showed cerebral signs, was restless and developed neck stiffness. Lumbar puncture revealed protein of 200 mgm and slight leucocytosis. The following day a Schwartze operation on the right mastoid was performed. He showed improvement but two days later showed an abscess in the right axilla containing staphylococcus aureus. He was eventually discharged from hospital at the age of $3\frac{1}{2}$ months and seemed well. At the age of 5 months he began to have convulsions. These were momentary and consisted of sudden attacks of pallor, rigidity and loss of consciousness but no definite twitching. The attacks persisted, occurring once a week for the next month and he was then admitted at the age of $6\frac{1}{2}$ months to a neurosurgical unit where hydrocephalic distension of the head, papilloedema and right hemipniesis were noted. A left frontal cerebral abscess containing staphyloccus aureus was aspirated and penicillin given. Fits persisted. They were noted to be focal in type with twitching commencing in the right thumb and fingers and face and gradually spreading to involve the whole of the right side. He was discharged at the age of 9 months, after 3 months in hospital and was recorded as having a right hemiparesis with hemianopia and focal epilepsy at this time. Regular phenobarbitone was prescribed.

His development was slow. He said 'Mamma'and'Dadda' shortly after the age of 1 year but other words were added only very slowly and even at the age of 9 his speech is confined to imitative phrases, which he says readily and short phrases like "Ian want bathroom", "Go away" etc.

He sat up at the age of 18 months and stood shortly before the age of 3. He walked with support about the age of 3 and without support at the age of 4. When he began to walk he was very unsteady and was always falling and banging into things on his right hand side. He still tends to bump into things when excited and has difficulty going down stairs and on rough ground.

He rapidly used his left hand after discharge from hospital but the right hand was never used until he was 6 or 7 years old, and then only to help the left.

Following /

Following discharge from hospital he had no fits for 3 months but then in the space of a few hours had 5. His breathing became heavy, face flushed, and the limbs on the right became tightly flexed. There was impairment but no loss of consciousness. After a few seconds he appeared drowsy but otherwise normal. Attacks similar to these have continued to occur 3 or 4 times in as many hours or days at intervals of 2-3 months. In the past 2 years, however, the contraction of the right limbs during the episodes has become less marked and been replaced to some extent by severe twitching of the arm and leg.

Between the age of 1 year and the age of 3 or 4 a gradual change took place in his behaviour. Previously he had been sleepy and lethargic but gradually he became more active and interested, "busy with himself". As his walking improved his activity became definite overactivity and he became irritable, rapidly roused to temper, destructive and unmaneagable; paid no attention to what was said and appeared to understand very little of it, though responsive to an affectionate or scolding tone of voice. He has no power of concentration and is incapable of playing with or being interested in anything for very long. He sucks his thumb a great deal and from the age of 4 has had a piece of rope which he drags around with him and sucks. He also sucks his clothes. He sleeps well.

Examination. Impossible to make accurate measurements on account of uncontrollable and violent behaviour both to me and to the mother.

Head circumference 21". Right arm approximately 1" shorter than the left.

His behaviour was grossly abnormal. He was overactive, never still. His attention was fleeting and he three any object that interested him about in a meaningless fashion. The objects that interested him were shiny or bright in colour. He appeared to understand very little of what was said but knew the words "bring" and "take" and the names of a few common household objects. He obeyed a few simple commands, but not consistently. He understood more speech than he could utter. His vocabulary was very poor and limited, yet he was capable of imitating very complicated sentences, even in French, though these were meaningless to him. His mental functions appeared to be very poor. The most complicated thing he did was to take chocolate to his mother for unrapping.

Complete right homonymous hemianopia on confrontation. Eye movements full.

Right facial paresis of moderately severe degree and upper motor neurone type.

The /

The power of the right limbs is much less than that of the left and the difference is more marked in the arm than the leg. Co-ordination in leg and arm is more impaired than power. Voluntary supination of the forearm and extension of the elbow are severely limited though passive movements are limited to a lesser degree. Finger extension is limited voluntarily markedly, is very weak and co-ordination of these movements is very poor. He can grasp and hold objects in the right hand but cannot release them. There is greatly increased tone of the right limbs. The increase is spastic in type and the biceps, triceps, supinator, knee and ankle jerks are much increased. The plantar response was extensor on the right and flexor on the left. The gait is abnormal. He walks on the right toes and does not put the heel to the ground. He brings the foot round in a circle when he walks. When he walks or used the left arm there is a marked tendency for the right to assume a position of flexion at the elbow, pronation at the forearm and clenching of the fist over the right thumb. It was impossible to produce elbow extension on the right as an associated movement by means of the tonic neck reflex.

Sensory findings untestable but mother does not think that he feels so well with the right hand as with the left.

The right hand and foot are colder than the left and there is some cyanosis of the skin over each.

Involvement of other cranial nerves.

The eyes.

Slight asymmetry of the pupils and reactions to light and accommodation being somewhat brisker on one side than the other were encountered not infrequently in cases of hemiplegia, without apparent visual defect or other evidence of oculomotor involvement. This finding is in accordance with the old observations of Koenig (1896).

Strabismus occurred relatively commonly in cases of hemiplegia in childhood and limitation of eye movements was noted in 22 cases. In 13 of these one eye was affected,

in /

TABLE XIb.

Strabismus in hemiplegia

Type of hemiplegia	Number of cases	Unilateral involvement		Bilateral Involvement		Total
		Oculomotor	Abducent	Oculomotor	Abducent	
Mild	27	0	3	0	1	4
Moderately severe	26	1	4	1	6	12
Severe	22	1	4	0	1	6
Totals	75	2	11	1	8	22

in 9 both eyes. In three cases the oculomotor nerve was affected, in 19 cases abduction of the eyes was limited. No duductions were possible about the aetiology of the strabismus. It was stated to have been present in a number of other patients during infancy but had gradually become less marked and was absent at the time of the examination. Most of the cases of strabismus tended to show improvement during childhood, and the majority in which abduction of the eyes were limited appeared to be of neuromuscular rather than central origin. There was no greater incidence of strabismus in the eye on the affected, than on the unaffected side. Table XIb.

Facial palsy.

Facial palsy was stated to be uncommon in hemiplegia in childhood in most of the earlier reviews. (Osler, 1889; Lovett, 1888). In this and other recent series, however, it has been found to be present, at least in latent form in the majority of cases.

The form of the paralysis varies somewhat in type and greatly in severity in different cases as noted by Koenig (1896) and Freud (1897).

The various forms encountered are indicated in Table XIc. Though the facial paresis tended to be more apparent in cases with moderately severe or severe palsy of the limbs a number of mild cases showed quite marked facial involvement.

It will be seen that 14 cases showed no abnormality of facial /

TABLE XIc.

The facial paresis in 75 cases of hemiplegia

Type of facial paresis	Number of cases
No abnormality evident	14
Facial asymmetry at rest and on movement.	21
Face normal at rest but lagging on the affected side on all movements.	31
Facial asymmetry apparent only on voluntary movement, not on emotional movement, or at rest.	4
Face lags on voluntary movement but overacts on emotional movement. Symmetrical at rest.	5
Total	75

facial movements. 21 cases showed asymmetry of the face at rest and on movement 31 appeared to show facial symmetry at rest but on movement, whether emotional or voluntary in type, the affected side moved less than the normal and asymmetry was produced. A smaller proportion of cases showed very interesting types of abnormality. In five cases the face was symmetrical at rest but on movement of voluntary type the affected side showed definite lag. On movement of emotional type, however, the affected side showed overaction, and the asymmetry appeared, literally, to be on the other cheek. Four cases showed symmetry at rest, and on emotional movements. But on voluntary movements a lag was present. The types of facial paresis encountered are similar to those described by Freud and Rie, (1891) except that he found a larger proportion of cases showed overaction on emotional movements. In his series approximately the same proportion of cases, 70%, as in this series showed little or no asymmetry of the face at rest.

Hearing.

A small number of cases showed middle ear deafness due to chronic otitis media with or without mastoiditis, which had indirectly led to the hemiplegia in a number of cases. One case whose pneumococcal meningitis had been treated with streptomycin showed severe, but not complete nerve deafness.

Two /

Two of the cases found to be aphasic had been diagnosed previously as being deaf.

Swallowing.

The frequent drooling seen in infancy in cases of hemiplegia appears to be due in most cases to infrequent swallowing. It is more common in mentally defective children than in others. It tends to improve at about the same age as the child learns to speak.

When the frequency of swallowing is noted in drooling children it is quite frequently found that they swallow as infrequently as once in two and a half or three minutes. Clearly this phenomenon may be due to the mental inertia of the child or to true difficulty in swallowing. In three cases, aged 9, 3 and 3½ respectively, all with great speech difficulty and showing drooling, swallowing of solid food was difficult and the palatal reflexes were abnormal, sluggish in one case and asymmetrical in two. Only one of these children appeared to be severely mentally defective.

Though relatively few cases with true difficulty in swallowing were found it seems possible that a number of cases of congenital hemiplegia do in fact have definite neurological cause for their difficulty in swallowing, and therefore, feeding in their first few months of life.

Shoulder movements.

Two cases were encountered in whom there was weakness of voluntary head rotation away from the side of /

of the lesion. One of these cases showed some winging of the scapulae. One was classified as a moderately severe case of limb paresis and showed facial, tongue and speech involvement. The other was a severe case of limb paresis without any other evidence of cranial nerve involvement but with definite dwarfing of the trunk on the affected side.

Tongue involvement.

Involvement of the tongue was encountered much more frequently than expected and was present in 9 cases. In three involuntary movements of the tongue were evident when it was produced. In one case these were severe and appeared to be generalised. In the other two cases they were slight and confined to the side of the paresis. Only the case with generalised involuntary movement showed speech defect.

In six cases some weakness of the tongue was evident on voluntary movement and the tongue tended to be deviated to the paretic side when central protrusion was attempted. Two of the six cases showed marked speech defect, the speech of the other four was easily comprehensible. Of the cases showing paresis of the tongue all were classified as severe or moderately severe cases of hemiplegia.

Head asymmetry.

The fronto-occipital circumference and hemicircumferences were measured in all those in whom it was possible /

possible to obtain reasonably accurate measurements. The skull was found to be asymmetrical by ½" or more on measurement in 9 cases though it was plagiocephalic in a much larger number. Only two patients with acquired hemiplegia showed a difference in the measurements of the affected side of the cranium and the unaffected, whereas 7 cases of congenital or probably congenital origin did so. The commonest asymmetry was a flattening of the patiental or patiental and occipital regions on the side of the head opposite to the paralysis. This was not always marked enough unfortunately to produce any significant difference on measurement, however striking to the eye.

Variations in the head circumference in hemiplegic and normal children, and the incidence of various head shapes will not be discussed as measurements of controls suitable for comparison are not yet available.

Trunk asymmetry.

Unfortunately routine measurements to determine trunk symmetry were not made though the trunk was measured bilaterally in most cases showing marked shortening of the limbs. In only three cases was the chest hemicircumference found to be smaller by ½" or more on the paralysed side. Two of these cases were severe and one moderately severe cases of paresis. All showed /

showed marked limb shortening on the affected side.

Epilepsy in hemiplegia in childhood.

The earlier authors were greatly interested in the relationship of epilepsy to hemiplegia in childhood. Some considered epilepsy as a frequent cause of hemiplegia, others as a symptom of the same underlying disorder as caused the hemiplegia. Some considered that it was symptomatic on the hemiplegia itself. The early work was reviewed and several important contributions were made to the study of epilepsy in hemiplegic paralysis by Wuillamier, (1882). He defined three periods in the development of epilepsy in acquired hemiplegia. The first stage consisted of convulsions occurring at the time of the appearance of the cerebral abnormality, most commonly during the course of infectious disease following birth. The stage of convulsions with its accompanying acute illness was succeeded by the period during which the hemiplegic paralysis developed. In most cases the convulsive attacks were in abeyance during this stage. In some they disappeared completely, but in most, epileptic attacks recurred after a period of time. This period of ti e varied greatly. In some patients it was as long as 18 years, but in others there was no period during which convulsive attacks were absent, the first convulsive /

convulsive stage merged into the third epileptic stage. In the majority of cases, however, convulsive attacks were absent for from one to three years.

Broadly speaking the conclusions that Wuillamier reached about the occurrence and nature of epileptic phenomena in hemiplegic paralysis were accepted and have been found to be true for the majority of cases in this series. All but three of the acquired cases of hemiplegia showed convulsive attacks in the acute stage of the initial illness. The large proportion of congenital cases showed convulsive attacks in the first few days of life. The nature of the initial convulsions varied greatly. Some showed generalised clonic and tonic manifestations, but the majority showed the most marked manifestations on the side later found to be paralysed. In most cases the fit began in the hemiplegic side and though in most cases after the preliminary convulsive movement the fit became generalised, clonic movements were usually more marked on that side throughout the fit. The severity and the duration of the initial fits varied greatly.

As the acute stage settled and the hemiplegia became apparent the fits ceased in the majority of cases, but they persisted in 5 cases without remission. In the remaining cases they were absent for variable periods of time, but then, convulsive attacks, not necessarily of similar type to those initially present, recurred.

Chronic epilepsy, the end result of the former

TABLE XId.

Epilepsy in 75 catients with hemiplegia in childhood.

Severity of hemiplegia	Number of cases	Type of attack			No. of patients with epilepsy (6 with more than 1 type of attack)
		Grand mal.	Jacksonian	Petit mal.	
Mild	27	4	2	0	5
Moderately severe.	26	7	3	4	12
Severe	22	4	4	0	7
	75	15	9	4	24

series of events was present in 24 cases of the 75 cases of hemiplegia in the series, as shown in Table XId. It will be seen that 4 cases showed more than one type of epileptic attack. Three showed petit mal and grand mal and one showed petit mal associated with Jacksonian attacks. The latent periods following the appearance of the hemiplegia during which no epileptic attacks occurred are shown in Table XIe.. It will be seen that the interval was less than two years in the majority of cases, and in all of whom the only epileptic disturbance was petit mal.

Three main forms of epileptic attack occurred though variations of these were not uncommon. The commonest was grand mal. Some of these cases showed the most marked clonic movements during the attacks on the side of the paralysis but in all the attacks were generalised, rath rathan localised, and began suddenly, not being confined initially to one limb. The next most frequent form was Jacksonian in type, the first abnormal movements occurring in the affected limbs and generally because consciousness was slowly lost or not lost the child could localise its origin precisely.

Two cases with Jacksonian attack were of especial interest, one showing predominantly sensory abnormalities in the initial stage of her attacks, and the other, previously quoted experiencing visual hallucinations.

The /

TABLE XIe.

Period of freedom from fits in 24 hemiplegia children with epilepsy

	No. of cases	No freedom	Less than 1 year	Over year but less than 2 yrs.	2 yrs. but less than 3 yrs.	3 yrs. or more
Grand mal	15	2	3	6	1	3
Jacksonian attacks.	9	3	1	2	3	0
Petit mal only.	4	0	4	0	0	0
	24 x	5	8	8	4	3

x 4 patients with attacks of more than 1 type.

TABLE XIg.

Effect of therapy on frequency of attacks in 24 hemiplegic patients with epilepsy.

Type of attack	No. of cases	Having therapy	Attacks less frequent on therapy.
Grand mal	15	12	10 x
Jacksonion attacks	9	6	4
Petit mal	4	2	2

x including 2 with no attacks for 2 years on therapy

The severity of the attacks and the frequency of their occurrence varied greatly. In Jacksonian attacks loss of consciousness tended to be more gradual, less profound and attacks were accompanied by more premonitary symptoms than were found in attacks of grand mal type. This was of obvious importance from the point of view of the child recognising his attacks and being able to put himself into conditions of relative safety before they were fully developed.

The frequency of the attacks of each type is shown in Table XIf.. That epileptic attacks appeared to be commoner in those with moderately severe than either slight or severe hemiplegia is shown in Table . Possibly the severe cases had more destroyed and less damaged cerebral tissue than had the moderately severe cases.

One point of some clinical interest was that in some patients the hemiplegia was apparently more severe following severe epileptic attacks for as long as two weeks after these had ceased, a matter of obvious importance so far as limb function was concerned.

As shown in Table XIg. the majority of patients with epileptic attacks received anti epileptic drug therapy. Most patients received several drugs alone or in combination at different times. They were successful in reducing the numbers of epileptic attacks in the majority /

TABLE XIf.

Frequency of epileptic attacks in 24 children with hemiplegia and epilepsy.

Type of epilepsy	No. of cases	More than one per day	More than one per week	Less than one per week More than one per month	Less than one per month More than one per year
Grand mal	15	0	4	3	8
Jacksonian attack	9	0	7	1	1
Petit mal	4	4	0	0	0
	24 x	4	11	4	9

x 4 had more than one form of epileptic attack

Behaviour disorder in hemiplegia.

Functional behaviour disorders occurred commonly in patients with cerebral palsy of all types. They were most frequently due to the child feeling frustrated as a result of his physical disability and to him comparing his performance unfavourable with that of other children, and especially younger siblings. Others were unable to adapt to society because of the excessive attention and over protection which they received in their early years.

The most common disorder, not apparently of functional origin, was that the child showed escessive disordered activity which appeared to be compulsive in type. The behaviour of these children was remarkably similar in all the cases in whom it was found. The child was never still. He handled everything in sight, and played with it actively, but in a facile manner, not showing much intelligence as he did so. His attention span was extremely poor and no object held his attention for more than a few seconds. Concentration seemed to be completely lacking in most of these patients. Their intelligence could not be fully assessed and of the 9 cases encountered, 6 of the more severely affected were classified as ineducable or under the age of 5 and likely to be so. Four of the cases were epileptic, 2 showed hemianopia. Seven of the cases were of acquired hemiplegia and the histories in all were remarkably similar. Following the acute /

acute illness during which the hemiplegia was acquired the child was quieter and more passive in behaviour than before his illness for a period of some months, but seemed to be somewhat out of touch with his surroundings.

Gradually he appeared to show more interest in his surroundings and at the same time, as his muscular functions improved, became more active, and eventually hyperactive. The child appeared to live in a world of his own, paying little attention to relatives and requiring only ministrations to animal needs. As his hyperactivity increased he became quite uncontrollable and any new object demanded to be handled, and is dif icult to handle, destroyed. Sleep is normal in most cases but little of it is required and the child's fund of energy appears to be inexhaustable. After a period of months or years the child's hyperactivity becomes less; he becomes more amenable to discipline and in favourable cases education, to a limited degree may be possible.

Case 144. Male. Born 1948.

Left hemiplegia, epilepsy, overactivity and mental deficiency following cerebral thrombophlebitis.

Born to healthy parents. Mother left handed. He was the second child, the first, a boy being born after a normal pregnancy and delivery is normal, but ambidextrous.

She was well in her second pregnancy with the patient until the last month when she developed severe pelvic pain. She was delivered at home spontaneously without difficulty. The child cried at once and the birth weight was 7 lbs. The neonatal period was normal. At the age of 4 months the child developed a discharging right ear. In spite of treatment the ear continued to discharge for a matter of months. /

months. The child sat with support at the age of 7 months. He stood at the age of 15 months and walked without support almost immediately afterwards. He spoke his first words at the age of 15 months but was rather slow to add words so that he still had no sentences by the age of 22 months. All the milestones were a month or six weeks later than those of the first born. From the age of about 4 months he used both hands to play with toys and shortly after a year began to use the left hand more than the right. Mother thought he was going to be left handed.

At the age of 22 months he developed a head cold. The right ear discharge increased and after one week of fever, listlessness and anorexia associated with the catarrh he had a convulsion with sudden loss of consciousness and generalised twitching of all limbs. He was admitted to hospital and was noted to show generalised increase of muscle tone, fever, a discharging right ear, positive Babinski signs bilaterally, and normal lumbar puncture findings. During the first three days of his stay in hospital a left hemiplegic paralysis became defined and a right jugular thrombosis was noted. Fever persisted and he was transferred to a neurosurgical unit. He was treated with streptomycin and heparin and 5 weeks after his convulsion the fever settled finally and he was discharged back to the original hospital where he developed whooping cough followed by measles. He was eventually discharged home 6 months after his convulsion. At this time he was just beginning to stand again. He was miserable, lethargic and tended to sit or lie whining most of the day. He could say nothing and used the left hand hardly at all. The mother noted that there was a squint, not evident before his admission to hospital. When he walked he tended to drag the left leg. His appetite was good and he soon showed a marked tendency to put anything he handled into his mouth and chew his clothes. Gradually over the course of a year a change took place in his behaviour. On discharge from hospital he had tended to be drowsy and slow. He slept a great deal and noted little but as time passed he began to wander about more and more, handled everything in sight and put them into his mouth. He became progressively destructive, tearing and eating anything he could reach. He did not seem to be able to comprehend anything that was said to him and attempts to control him seemed to make his destructive behaviour worse. 18 months after his discharge from hospital he did seem to begin to understand simple commands.

At the time of his return from hospital he had fits every day or every other day. These started gradually by his going pale, the eyes glazed and he would lose consciousness for about 10 minutes during which time the left side only showed clonic movements of the limbs. After recovery of consciousness he /

Case I44. Patient with acquired right hemiplegia, hemianopia and gross overactivity, apparently oblivious of his surroundings.

he would sleep for several hours. Since his discharge from hospital the fits have gradually become less frequent and less prolonged and in the past 18 months last no more than 5 minutes, are accompanied by a tonic state of the left limbs but no clonus and occur only once a week or so. His behaviour has remained unaltered and he still has no speech. He uses the left arm only when he has to and only to assist the right.

Examination. Height 41¼". H.C. 20½". There is ½" smaller measurement on the right than the left hemicurcumference of the skull. There was 1/3" shortening in the left arm and ¼" shortening in the left leg compared to the right.

The child was grossly overactive in behaviour. He was never still. He fiddled with objects continually and chewed anything in sight. He paid no attention to or could not comprehend commands. He shrieked but had no speech, and could not imitate. There was a bilateral paresis of the sixth nerves, of severe degree. The face showed a weakness of the left side compared to the right. There was a left paresis of the upper motor neurone type.

He was very strong but the left limbs were less powerful and less well co-ordinate than the right. There was a few degrees limitation of supination of the left forearm and of the extension at the left elbow passively. Otherwise passive movements were full. In the left leg there was limitation of dorsiflexion of the ankle. The extent of voluntary movements could not be tested, but appeared to be little limited.

When made to move the left hand the finger movements were grossly inco-ordinate and there was marked slow separation and extension athetosis involving all the fingers and the wrist. He could grasp objects but not, apparently, release them.

The gait was grossly impaired with the left foot in a position of marked equinus and the hip tended to adduct so that the left foot was placed almost exactly in front and across the right. As he walked or when he used the right arm the left took up a position of adduction at the shoulder, flexion at the elbow, wrist and fingers and pronation of the forearm. There was a moderate increase of tone of the left limbs which were spastic. The biceps, triceps, supinator, knee and ankle jerks were much increased on the left compared to the left. There was no clonus. The Hoffmann signs were absent. The left plantar response was extensor, the right flexor.

Sensory findings untestable except that pin prick was appreciated on the left limbs.

The /

Case I7.Patient with right hemiplegia,hemianopia and gross overactivity.In his left hand he holds a piece of string which he is wont to chew.

The importance of this hyperactive disorder is that a number of these children have to be placed in institutions for their own and their siblings safety, and that in the majority, education is possible to only a very limited extent.

The type of hyperactivity is similar to that encountered in some epileptic patients (Peterson, 1953) and in some of these is apparently accentuated by phenobarbitone administration.

Educational difficulties.

Some of the specific educational difficulties encountered in hemiplegia will be considered in a later section.

Other abnormalities.

A number of other abnormalities of clinical interest though little importance from the functional point of view were encountered. A number of patients showed rather poorly developed hands and feet. These were small, and the fingers tended to be poorly differentiated, being short and tending to be similar in length. The fingers were frequently hyperextensible and syndactyly, especially in the toes was common.

Two patients had been born with bilateral talipes equino varus deformity and had later acquired hemiplegic paralysis.

The relatively high incidence of abnormality and poor development of the hands and feet in cerebral palsy was noted by a number of the earlier German and French authors, who suggested that they might be skeletal manifestations of general /

general faulty development, and possible indicate a greater susceptibility to neurological and other lesions. (Fere, 1896; Freud, 1897).

It is not proposed to discuss these findings in more detail as they are more in the nature of aetiological than functional problems. But the high incidence of syndactyly and epilepsy in the parents of children with hemiplegic paralysis was interesting.

One patient had a congenital dislocation of the hip on the hemiplegic side which had defied all orthopoedic attempts to reduce it.

The clinical findings in 79 cases of diplegia.

By diplegis is understood a condition of more or less symmetrical paresis worse in the legs than the arms and dating from birth or shortly afterwards. (Freud 1893). The The paresis is accompanied by rigidity or spasticity in the affected limbs in most, but not all, cases. The condition is very often associated with mental defect, epilepsy and strabismus. More than one third of the children with diplegia are born prematurely. Abnormal pregnancy, especially ante-partum haemorrhage and pre-eclamptic toxaemia, abnormal delivery and neonatal asphyxia are frequent antecedents of diplegic paresis. It was only rarely a postnatally acquired condition. The term diplegia is used to refer to types of cases so called by Freud, but excludes his categories of bilateral hemiplegia and athetosis and chorea.

The course of diplegia.

The majority of cases of diplegia have their origin before, or at the time of birth, but it is usual for there to be a delay of some months in the appearance of the stage of diplegia marked by rigidity or spasticity. The majority of cases are only recognised by doctors and clinics when this final stage of spasticity or rigidity has appeared. Yet the sequence of events from the time of birth until this stage has appeared is very typical and its study leads to a much greater understanding of some of the more bizarre phenomena found in the later stage.

Unfortunately, owing to the ignorance of the early authors /

authors so far as the new born period was concerned, little attention has been paid to the earlier stages of the development of diplegic paralysis, though a few authors realised that these were marked by more than rigid or spastic paralysis of slight degree. (Little, 1843; Osler, 1889; Parrot, 1873). Since much of the later interest in the condition has come from orthopoedic surgeons interested in the overcoming of deformities rather than their mode of development, the earlier stages have remained rather neglected, though at least one modern system of treatment owes much to their study. (Bobarth and Bobarth 1950).

The hypotonic stage of diplegia.

In the first few weeks after birth the majority of cases of diplegia show little apparent abnormality. Even on examination the child may appear to be quite normal, as is attested by the numbers of children in the present series who were thought to be normal by infant clinics and later developed diplegic paralysis. In 31 cases, however, a story was elicited from the mother, of the child being floppy in the weeks after birth. In a few cases definite hypotonia had been noted by infant clinics and doctors. "His head rolled all over the place", "He felt like a half filled pillow", are typical descriptions offered by the mothers of these children. Another early deviation from the normal is a certain poverty of movement. The child is noted to move less /

less when placed to rest or when stimulated, than a normal child. "You'd lay him down and he was just the same in the morning; you'd think he hadn't breathed all night".

This stage of hypotonia persists for a variable time in different children. It disappears rather abruptly with the appearance of the first signs of postural activity, and especially the first attempts to hold up the head. The age at which this occurs is variable.

The dystonic phase.

With the first attempts at holding up the head the stage of apparent normality or, more accurately, little apparent abnormality, usually ends. The mother finds that when she changes the child's position abruptly as she handles him, usually bathing or feeding, the child suddenly becomes stiff in her arms. He feels to be "all of a piece", (a phrase used by Little). The child's head is thrown backwards, the neck and back are extended. The upper limbs are adducted at the shoulders and slightly internally rotated. The elbows are extended, the forearms pronated and the wrists and fingers are usually flexed. The legs are extended at the hips and knees, and plantar flexed so that the toes point downwards. This position is achieved as a result of a sudden, extremely powerful dystonic movement involving the whole trunk and the limbs.

Once achieved the position is maintained for only one or two seconds and then the child relaxes and appears to be normal /

normal again. The dystonia is never accompanied by true loss of consciousness but seems to be associated with some impairment in some cases. The attacks may greatly distress the child.

Initially the attacks occur relatively infrequently, perhaps once or twice a day and they are elicited only by sudden and marked changes in position. In the course of one or two weeks, however, the attacks become more frequent and more readily elicited, so that they may occur 20-30 times a day or more. In one case it was sufficient to tap the sides of the cot to produce dystonia. The attacks are first noted when the child is being bathed, or when he is being dressed, but when they are occurring frequently attacks may be precipitated whenever the child is handled, however slightly. In some cases they appear as the head is being adjusted so that the mouth can take the nipple comfortably during feeding, and this may lead to feeding difficulty. The trigger for the production of dystonic attacks is extension of the neck or back in the early part of the dystonic phase. Sudden extension of the head, especially when the child is held upright will almost invariably produce an attack in children with dystonia. The attack resembles, very closely in the mode of its production and its manifestations the righting reflexes found in decerebrate animals. (Sherrington, 1905; Magnus, 1926). They can probably be regarded as the result of /

of postural reflexes which are not inhibited by cortical centres, and are, therfore, over-responsive and sensitive to changes in position.

The dystonic phase of diplegia perists for a variable time in different cases. It usually becomes less marked when the child achieves more perfect control of his postural mechanisms, and especially when he learns to hold the head steadily. When the child can prevent the head being extended abruptly by changes in position, the most potent trigger mechanism is no longer operative and attacks occur less often and require more severe stimulai to produce them. This stage of being able to control the position of the head usually co-incides with the beginnings of intelligent play with the hands, which is very variable in the age of its occurrence in children with cerebral palsy.

At the same time as the dystonic attacks become less frequent and less easily produced the first signs of stiffness in the limbs, commonly the legs, are noted by the mother. She may find that when she wants to place the napkin in position the legs are difficult to separate. She may have to pass the napkin between the legs on the end of a pencil. When she washes the legs they yield little to handling. She frequently observes that she has to open one or both hands with some force before she washes the palms, whereas before she did not have to do this.

<u>The stage of rigidity and spasticity</u>.

The /

The stage of rigidity of spasticity of the limbs emerges gradually from the dystonic phase. Initially the stiffness is first evident in the legs, especially the thighs, but gradually extends to involve the whole leg in paralegic cases and one or both arms in triplegic and tetraplegic cases. Initially the stiffness tends to maintain the limbs in positions similar to those seen in the dystonic attacks, positions of extension. The stage of extension lasts for a variable time; in some cases it persists for many years, but in the majority of cases it is succeeded, as attempts at voluntary activity become more frequent, by a stage of flexion of the limbs. Whereas the predominant increase of tone when the limbs tended to be extended was one of rigidity, when the limbs become stiff in flexion, the predominant increase of tone is spastic in type. The tendon jerks are slightly, if at all increased in the stage of extension, they are markedly increased in the stage of flexion. The Hoffmann and Babinski responses become positive.

During the earlier stages of rigidity in the limbs, voluntary movement tends to produce associated movements in the limbs which result in their being placed in extensor positions. Gradually however, in all but very severely affected cases positions of flexion of the limbs are assumed as a result of associated movements, and the change usually occurs as spasticity first becomes evident.

In patients in whom the more primitive extensor associated movements /

movements predominate over the flexor patterns, voluntary movements of the limbs are also more primitive in type. Thus in no cases in the present series in whom the extensor positions predominated, well well co-ordinated prehensile thumb and forefinger movements present. The movements of the hands were clumsy and infantile in type, the fingers being used together and opposed to the palm of the hand. The later developing thumb and forefinger movements appear only when flexor positions of the limbs are becoming predominant.

In tetraplegic cases in whom the limbs on one side are more severely affected than those on the other, associated movements tend to show extensor patterns for longer in the more affected side, than in the less affected. Sometimes a very marked difference in the manipulative ability between the hands is evident. The side with the more primitive movement pattern may show only clumsy grasping movements of the whole hand, whereas the side showing predominant flexor associated movements may show quite well developed thumb and forefinger movements.

Once flexor associated movement had spastic increase of tone present in the limbs, the complications of voluntary movement begin to bear more similarities to those found and previously described as occurring in hemiplegic patients.

Athetosis, of a similar type to that seen in cases of hemiplegia, is found in a large number of cases, especially those /

those with severely affected limbs in whom mobility of the limbs has been maintained. Movements of choreoid type occur less frequently.

Contracture, as in hemiplegic patients, is very liable to occur in limbs which are little used, whether because of severe paresis, or because of mental deficiency or lack of desire or need to move the limbs on the part of the child.

Two marked differences from hemiplegic patients may be noted, however. Marked vasomotor abnormalities are uncommon and dwarfing of the limbs is only exceptionally asymmetrical.

The relationship of the stages of diplegia to each other.

The typical case of diplegia of moderately severe or severe degree exhibits, at different times all the stages of the condition, hypotonic, dystonic, rigid and spastic and they eventually show generalised spasticity with contracture. All these stages are evident in most cases of diplegia in which the arms as well as the legs are affected. In younger patients a history of the hypotonic and dystonic phases of the condition was obtained more often than in the older patients, presumably because the temporary phenomena of these early stages had been forgotten in the latter group.

It was found that more severely and extensively affected children tended to show hyptonia and dystonia for longer than less affected patients. In patients with mild

triplegia /

triplegia and paraplegic paresis a history of hypotonia and dystonia was frequently not obtained, either because they were so mild that they had not been observed or because they had not occurred. On the other hand, very severely affected tetraplegic cases tended to show manifestations of dystonia long after the true dystonic attacks had disappeared. These manifestations were most commonly observed when the child began to walk. The combination of pressure on the soles of the feet, attempts to balance the body and to perform reciprocal movements, was enough to produce paroxysmal involuntary extensor movements of the head, neck, upper trunk and limbs. These paroxysmal movements, in most cases only produced on massive bodily activity, are the origin of the extensor thrust which such cases show. In these patients voluntary movements involving much associated postural activity, tended to produce positions of extension in the upper limbs, instead of the more usual flexor associated movements. In a few cases these extensor movements were sufficient to disorganise all such attempts at voluntary movements. In them it was possible to elicit modified dystonic movements experimentally. Thus the division into hypotonic, dystonic, rigid and spastic phases of diplegic cases is somewhat arbitrary. A number of children whose predominant disorder is of rigid or spastic type, will be found to show well marked dystonic phenomena.

A small number of patients with moderately severe or severe /

severe diplegia with triplegic or tetraplegic involvement apparently show no dystonic phase. A much larger proportion of less severely affected cases of paraplegic distribution show no dystonic phase, though frequently dystonic attacks may be produced in these children, experimentally. A few cases appear to miss one or more of the phases. In them the hypotonic stage may be followed directly by the appearance of spasticity in the limbs, or rigidity may be noted within one or two weeks of birth. In no case, however, has dystonia been noted to follow spasticity, or hypotonia rigidity.

During the discussion of the different stages of diplegia no ages for their appearance have been given. This is because of the marked variation that is found in different cases. Thus if a child with severe diplegia and mental retardation makes no effort to hold up the head until the age of 15 months, then dystonia is unlikely to occur until that time. Until the age of 15 months he will be in the hypotonic stage, floppy, showing poverty of movement and apparent lethargy. Once dystonia has appeared it is unlikely to become less marked until the head can be held more or less steadily and the hands are being used a little. In one case in the series this was not until the age of 4. In the majority of diplegic children in whom attempts at holding the head up, holding it steady, and using the hands are only slightly later than in the normal child, the stages tend to appear at similar ages. The age at which each stage appears is related to the level of motor development /

TABLE XIII

The age at the onset of dystonic attacks and their duration in 79 patients with diplegia

Age	Before 2 months of age.	Over 2 mths. under 4 mths.	Over 4 mths. Under 6 mths.	Over 6 mths. under 1 year	Over 1 year
Duration					
Less than one mth.	0	0	1	0	0
More than one mth. less than 2 mths.	0	0	0	0	0
More than two mths. less than six mths.	0	5	3	1	0
More than six mths. under one year.	0	4	3	1	0
Over one year, under two years.	0	2	1	0	0
More than two years.	2	0	0	1	0
Present at time of examination.	1	0	3	0	2
Totals	3	11	11	3	2

development rather than to age. (Bobarth, 1953; Bobarth and Bobarth, 1950).

In children with triplegic or tetraplegic involvement in whom attempts are made to achieve these skills with little delay the dystonic stage commonly appears at the age of between 2 and 6 months and from two months to a year, before the rigid or spastic phase gains the ascendance.

It will be seen from Table that 4 patients had dystonic phases which persisted for more than one year and three for more than 2. In six patients the dystonic phase was still predominant at the time of examination. One of these cases aged 4 was showing marked decrease in the severity and frequency of dystonic attacks, after three and a half years in the dystonic phase.

Since, in general, cases showing persistent hypotonic or dystonic phases are more severely handicapped than those showing early rigid or spastic phases it seems logical to use the different stages as a basis for classifying types of diplegia encountered. In the following sections descriptions will be given of the various types of diplegic disorder found. An attempt will be made to show that many of the findings present in more severe diplegic cases and absent in those more slightly affected, are the result of the persistence of typical manifestations of the earlier stages of the condition.

The severity of diplegia.

Diplegic cases were classified, for convenience into categories /

TABLE XII

The severity of the paresis in 79 cases of diplegia in childhood.

	Mild	Moderately severe	Severe	Totals
Stage of diplegia				
Hypotonic	0	1	0	1
Dystonic	0	2	4	6
Rigid or spastic	12	33	27	72
Distribution of paresis				
Paraplegic	9	13	7	29
Triplegic	4	14	5	23
Tetraplegic	0	8	19	27
	13	35	31	79

categories of paraplegia, triplegia and tetraplegia, according to whether useful function of the legs only, the legs and one arm, or all the limbs was impaired. The categories were based on a study of functional impairment rather than on neurological findings, however. The differences were of degree and not of kind. In all but three cases labelled paraplegic, definite neurological abnormality of the upper limbs was apparent on careful examination. Impairment of the co-ordination of fine and rapid finger movements, impaired power of finger and wrist extension and of supination of the forearm were usually present. From the functional point of view, however, these findings were of relatively little importance in limiting the usefulness of the upper limbs.

Similarly cases called triplegic were all tetraplegic from the neurological point of view, but since the usefulness of only three limbs was impaired, they were classified as triplegic from the functional point of view.

The degree of impairment of function varied considerably, even in those with paralysis of similar extent. Thus paraplegic cases might be completely unable to walk or their gaits might be only slightly abnormal. It was considered to indicate the severity of the paresis, as well as its extent, as discussed in the section on classification.

The numbers of the different types of paresis encountered in the survey are indicated in Table XII. It will be observed that the numbers in the categories of paraplegic, triplegic and /

and tetraplegic paresis are roughly similar, but that the severity of the limb involvement tends to increase with the numbers of limbs involved. In tetraplegic patients it is more easily possible to observe the gradual change from dystonic to rigid and spastic stages and to account for some of their findings on the basis of the persistence of some of the dystonic phenomena into the rigid phase.

Cases in the hypotonic phase.

Since these patients are usually thought to be normal during the stage of hypotonia unless they are very mentally defective, few were seen in this stage of diplegia. All but one of those showing hypotonia who were seen showed early infrequent dystonic attacks and were, therefore, placed in the category of dystonia. The exception was a boy of two and a half years who had been hypotonic from birth and in whom very early spasticity of the limbs was present. At no time had he shown any dystonic phenomena. His motor control was extremely poor though he could sit without support. His hands were used in a very infantile fashion. There was slight bilateral athetosis of the fingers. He was somewhat mentally defective. He showed marked hypotonicity on handling but his tendon jerks were increased in all limbs and the plantar responses were bilaterally extensor.

As might be expected from the fact that cases showing hypotonic diplegia are showing the most primitive form of motor /

motor defect, many of the children are said to die as a result of infections and progressive cerebral deterioration in early life. (Phelps, 1941).

The sex distribution and extent of the paresis in the diplegic patients.

Thirty of the diplegic patients were females and 49 were males. The differences in the sex distribution in the various types of paresis, paraplegic, triplegic or tetraplegic were not significant.

It will be noted that there was an approximately equal number of patients showing paraplegic, triplegic and tetraplegic paresis.

These figures are roughly similar to those of Freud and the majority of modern series. Freud quoted a series of 69 cases of which 30 were of paraplegic distribution and 39 triplegic or tetraplegic. The only difference between this series and most modern series is that the latter tend to include a rather higher proportion of triplegic cases. Hillebrandt, 1951; Asher and Schonell, 1950). Obviously differences in the criteria used in classification and the nature of the cases included in the various series could account for these relatively small variations in the relative frequency of each form of paresis.

Six cases were in the dystonic stage of diplegia when they were examined and it was possible to observe the transition of three of these cases to the stage of rigidity within a period of months.

Two of the cases were triplegic, four were tetraplegic; all were considered to be severe or moderately severe cases of diplegia. All the children were under the age of four years and three of them were under the age of 12 months. None of them was able to sit during the stage of dystonia and manual manipulation was very infantile and clumsy.

One case was observed first in the stage of hypotonia. Dystonia commenced at the age of 11 months and was still present at the age of 2 years three months. By this time some rigidity of the legs was evident.

Case 13. Male. born 1952.

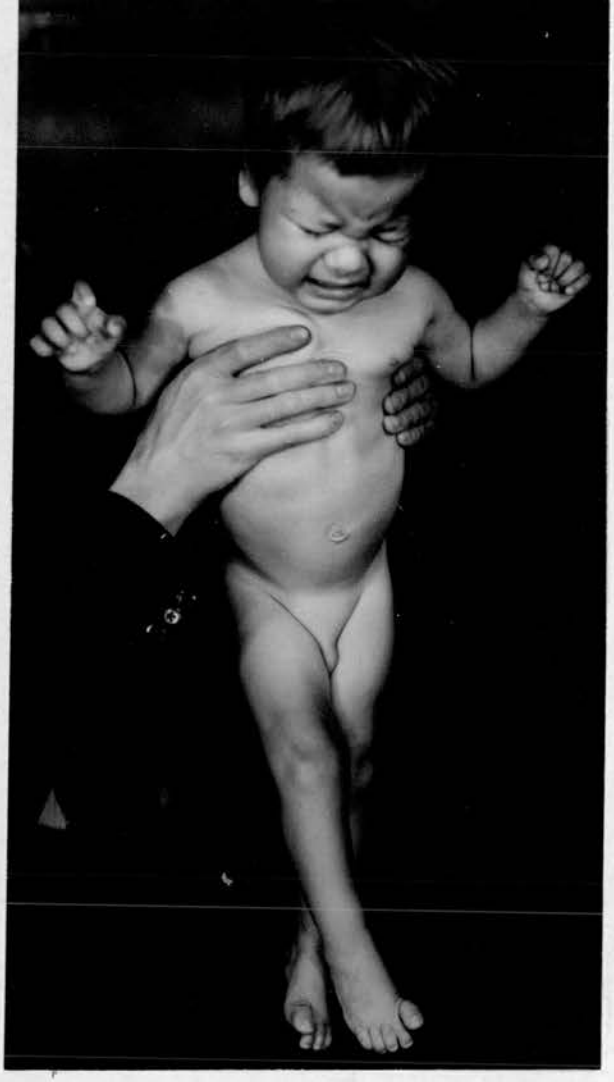
Severe dystonic tetraplegia, mental defect and retrolental fibroplasia.

The second of two children born to mother aged 22 and her husband. Both parents tuberculous.

All the family right handed. The first child was born at term after normal pregnancy and delivery, and is well.

In her second pregnancy she was extremely worried because her husband was ill and unemployed. She felt tired and had recurrent colds.

Nine weeks before term she had a profuse vaginal haemorrhage. She was admitted to hospital and the child was delivered spontaneously under chloroform analgesia after a labour of $5\frac{3}{4}$ hours. The child was cyanosed at birth and would not breathe, but responded to oxygen and injections in a /



Case I3.The dystonic stage of diplegia.In the upper picture the childs inability to hold the head is shown. In the lower the position resulting when the head is briskly extended,is shown.

a matter of minutes. The weight was 3 lbs. 8 ozs. The placenta was pale and weighed 15 ozs.

The child was placed in an incubator and was in continuous oxygen for 3 weeks. At the end of this time the child developed a respiratory infection which necessitated penicillin and later blood transfusion. He was discharged at the age of 11 weeks when the birth weight was 6 lbs. 8½ ozs. It was noted at the time of discharge that the child did not appear to be seeing yet and that there was persistent nasal catarrh.

At the age of 15 weeks the child was admitted to hospital with pneumonia and in the first year of life received antibiotics in hospital and at hone on seven occasions on account of otitis media, respiratory infections and bronchopneumonia. There was persistent chronic nasal catarrh.

When the child was discharged the mother noted that he felt "floppy" compared to her first child. He fed slowly, was windy and at the age of 6 months the mother suspected he was backward because he was not attempting to hold up the head and not looking about. He held up the head at the age of 8½ months, at which time bilateral retrolental fibroplasia was diagnosed. At the age of 10 months he was still not using the hands to play with toys, even when they were placed in them. The mother noted that when she washed him he frequently became stiff in her arms with the back arched, the head and legs thrown backwards, the forearms pronated, elbows extended and fingers semi-flexed. These dystonic attacks became more frequent and occurred on less stimulus, so that at the age of 1 year they were occurring many times a day whenever the child was handled. They were momentary only and the mother did not think there was any impairment of consciousness during them. The attacks persisted until the age of 2, but from the age of about 20 months they gradually became less frequent and occurred only when the child was bathed and not on handling unless this was vigorous. At the same time his legs began to feel stiff.

At the age of 2 the child was able to sit with support but had no independent sitting balance, no appreciation of speech, showed no recognition of its mother and appeared to be able to see only the difference between light and darkness. He appeared to be unable to swallow solids. Many exacerbations of his chronic catarrh had occurred with fever.

Examination. Length 28½". H.C. 18". Fontanelle still ½" patent at the age of 2. Occasionally smiles but otherwise very passive child resenting handling but showing no interest in its surroundings. Grunts and cries but no words. Much drooling. /

drolling. Able to hold up the head and sit with support. Moving the hands little, but the left more than the right.

Severe bilateral retrolental fibroplasia. Right pupil irregular. Both pupils reacted slowly to light. Bilateral abducent paresis.

Slight facial asymmetry due to right sided lag, on movement. Swallowing appeared to be infrequent.

There was weakness, stiffness and great inco-ordination of all limbs. This was very marked in the legs and the right arm but less marked in the left arm. There were no contractures, passive movements being full except for slight limitation of dorsiflexion of the feet bilaterally. There was a very severe increase of tone in the legs which were rigid and a slightly less severe increase in the right arm. The increase in the left arm was moderate. Stretch responses were present in all muscle groups in all the limbs. The biceps, triceps, supinator, knee and ankle jerks were much increased in all limbs, rather more brisk in the right arm than the left. The Babinski signs were absent but the Chaddock and the Oppenheim signs were bilaterally present. The hands were very soft and the fingers were gross hyperextensible, as were the toes. Postural reactions were marked, and interesting. When attempting to sit with support the child showed immediate flexion of the legs at the hips and knees and extension of the elbow with adduction and slight flexion at the shoulders, so that the arms were held rigidly and straight by the sides. When in this position the hands were tightly clasped. When the child was held in the horizontal position the arms were extended, as when sitting up but the legs were also fully extended. When the child was held under the arm pits the legs were extended and scissored. But at the same time the arms were adducted at the shoulders, flexed at the elbows, wrists and fingers and pronated at the forearms. The tonic neck reflexes were present bilaterally and the Moro response could be elicited inconstantly.

Sensory findings were untestable but light touch and pin prick were appreciated bilaterally. The hands and feet were cold to the touch.

Another case was seen shortly after the first attacks had occurred at the age of 6 weeks, at which time no rigidity was evident. The attacks had become very infrequent and much /

much milder by the age of 3 months, when the child was last seen, but the legs showed considerable rigidity and one arm slight rigidity by this time. Obviously such a short stage of dystonia is likely to be missed when mothers are questioned about it many years later.

Case 138. Male. 1952.

A case of severe triplegia in the dystonic phase.

An illegitimate child, born to healthy mother and father. Mother was aged 23 at the time of delivery. The patient was her first child.

She was well during pregnancy except that she was very large in the last 4 or 5 months of pregnancy on account of hydramnios. The labour was normal and lasted 18 hours. It occurred at term. The child was delivered spontaneously under nitrous oxide anaesthesis and appeared to be normal, crying at once. The weight was 8 lbs. 5½ ozs.

The child was discharged from hospital to a nursery for the purposes of adoption. He seemed rather floppy for the first three weeks but fed well and no other abnormality was noted.

Between the age of 3 and 4 weeks he began to have spasms in which he suddenly went stiff, three the head back and showed generalised rigidity and extension of the limbs. The attacks were momentary only and initially occurred most frequently when he was being bathed or washed. In the ensuing weeks he showed the attacks more frequently and at the same time they became more easily produced by less marked stimulation, especially feeding.

Examination at the age of 6 weeks. The child was alert and was making baby noises of pleasure and crying when annoyed. He could smile. There was a slight right abducent paresis and the face was asymmetrical because of weakness of the left face at rest and on movement. Swallowing was normal.

The child moved the right arm more than the left but neither as freely as a normal baby. The legs were little moved. The Moro response was present. When the child was held upright with the head flexed, the legs took up a position of slight flexion at the hips, extension at the knees and severe plantar flexion at the ankles. There was some scissoring in the thighs. The upper limbs tended to be flexed.

When /

When the child was held with the head extended and the head was briskly extended, immediate hyperextension of the neck and back resulted. The limbs were also extended, the upper limbs adducted at the shoulders, extended at the elbows and flexed at the wrist and fingers. The forearms were pronated. The lower limbs showed full extension at the hips, the knees were extended and there was a position of equinus of the feet bilaterally. The child was extremely rigid in this position. By jerking him suddenly so that head extension resulted, the opisthotonic attitude described above was immediately produced.

There was a suspicion of increase of tone in the left arm and a definite increase of tone in the legs bilaterally of rigid type. The tendon jerks were all slightly brisker than normal. Stretch responses could be elicited from both lower limbs and the left arm.

Examination at the age of 3 months showed some changes. The child was 24" in length, the head circumference was $14\frac{1}{2}$". There was a definite right abducent paresis and a slight left abducent paresis. There was a definite left sided facial paresis. The child could hear. Swallowing appeared to be normal.

The dystonic attacks could still be produced by extending the head rapidly. But they were less easily produced than on the previous examination. There was a generalised increase of tone in the lower limbs and in the left arm. The right arm showed a less marked increase of tone. The increase of tone was rigid in type. Stretch responses could be elicited in the left upper limbs and the lower limbs. The tendon jerks in these limbs were much exaggerated compared to those of the right upper limb. The Moro response was absent. The child showed a marked tendency to scissoring and extension of the lower limbs when held upright and the left upper limb showed a position of extension as an associated movement very readily. The right arm w as the only limb which was moved at all freely.

Sensory findings could not be tested in great detail but pin pricks appeared to be appreciated in all areas.

One case was seen in which dystonic phenomena persisted, in spite of increasing motor skill. In the previously quoted two cases it seems unlikely that dystonia was more than a transient /

transient phenomenon. In this case, however, it is likely that some dystonic phenomena will persist, for many years, if not for life.

Case 56. Female. Born 1949

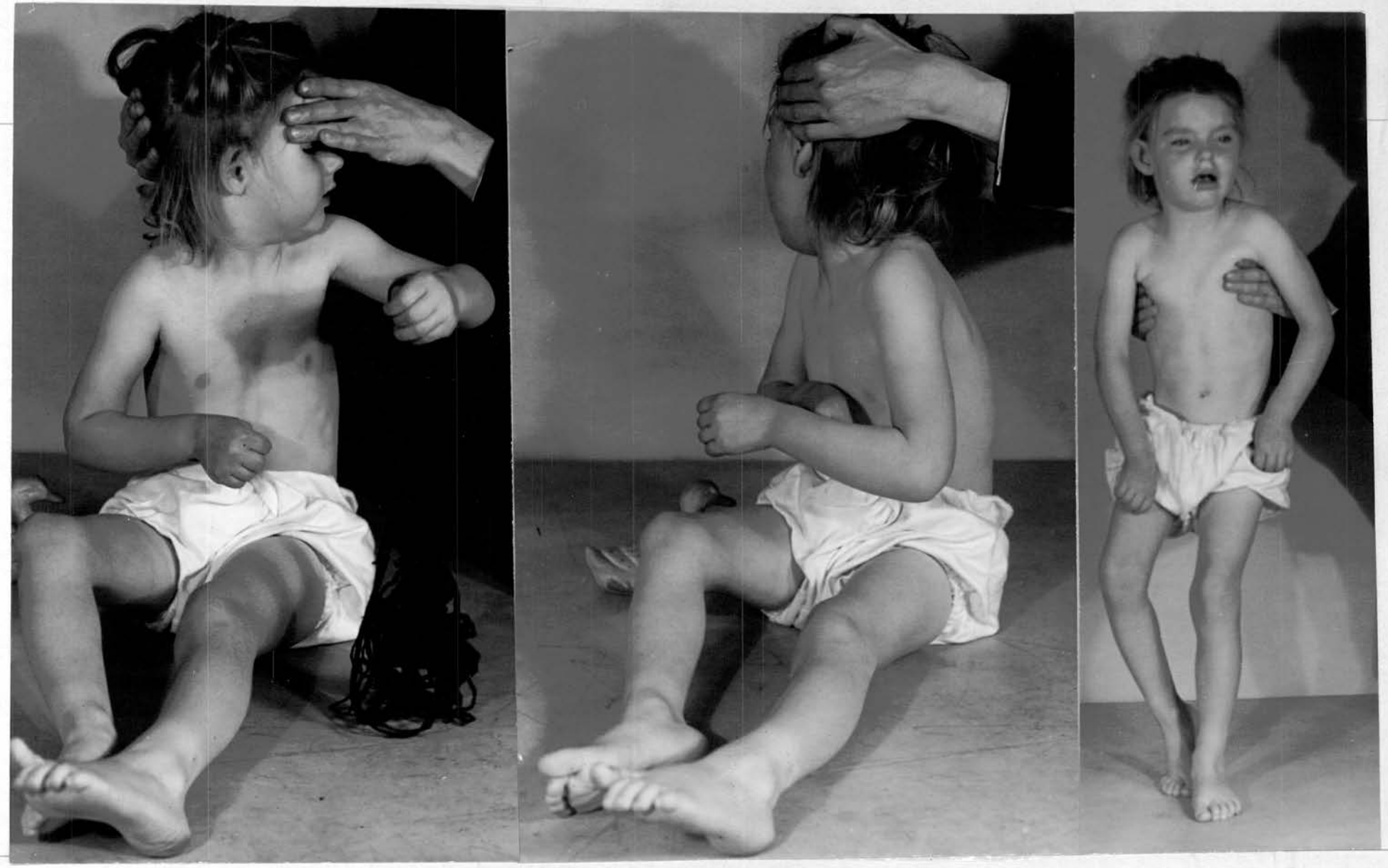
Severe dystonic diplegia, mental defect and epilepsy.

The first of two children born when the mother was 28. Both healthy and right handed. During her second pregnancy she was well, the child was born at term and is healthy. In her first pregnancy she had some vomiting in the first three months and was then well. The labour occurred 3 weeks after the expected time and, on account of primary inertia lasted 38 hours and 40 minutes before spontaneous delivery under chloroform analgesia was achieved. The placenta weighed 1 lb. 9 ozs. and showed gross infarction. The child weighed 6 lbs. 9 ozs. and cried at once following delivery. She was a quite infant, fed well and showed no initial abnormality.

At the age of 3 months the mother noted that though she was moving the left arm the right lay by the side with the hand clenched, fingers over the thumb and the palm facing backwards. She began to grasp with the left arm at the age of $4\frac{1}{2}$ months. At about this age the mother found that when she was handled or moved the left arm from the shoulder she was inclined to go stiff suddenly, the back arched, the legs extended, the arms adducted at the shoulder, extended at the shoulder, pronated at the forearms and flexed at the wrists and fingers. This position was assumed first when she was feeding, especially if she had trouble swallowing, and on washing. But it gradually became elicited on lesser stimulai and occurred 20 or 30 times a day, and persisted until the age of 3 having become steadily less frequent for about 6 months before that.

She held her head up slightly tilted to the right at the age of 3 months and it was steady by the age of 6 months. She could sit with support from the age of about 1 year and on a chair without support from the age of 20 months. She could sit on the floor from the age of $2\frac{1}{2}$. Could stand and walk a few steps with support from the age of 3. She said her first words at the age of 3 but appeared to comprehend speech from the age of $2\frac{1}{2}$.

At /



Case 56. Child in the late dystonic stage of diplegia. When there is no pressure on the soles of the feet the tone of the limbs is predominantly spastic and the associated movements are flexor in type. When there is pressure on the feet the limbs tend to be held in positions of extension and the tone is rigid.

At the age of 4½ - 5 months she began to have fits. These occurred suddenly and during them she would suddenly become limp and fall forwards, with impairment, but no loss of consciousness lasting about one second. Imeediately after an attack she appeared to be a little far away mentally, but after a minute or two, as before. The attacks varied in frequency from 3-4 a day to 20 a day, but after the age of 2 they gradually became less frequent and ceased at the age of 3.

She has remained clumsy and though she handles objects with both hands, knocks them over and drops them as often as picking them up and holding them successfully. Her left arm was always more used than the right and still is. The mother has noted that when she uses one hand the opposite arm seems to be in action too, and if she is energetic, tends to go stiff and straight, without her knowing.

Her behaviour has been very difficult. She is demanding, overactive and very easily frustrated. She showed affection for the first time at the age of 3½.

She had not achieved any bowel or bladder control at that time.

Examination. Height 39¼". H.C. 18½".

She was overactive and her behaviour was disordered. She appeared to comprehend a fair amount of what was said to her and could respond to simple commands. She responded to commands with requests for imitation than to commands alone. Her concentration was extremely poor and fleeting. She could laugh and shout but used no words during the examination.

There was slight bilateral abducent paresis.

The tongue movements were full but rather inco-ordinate and weak. Impairment of power and co-ordination was evident in all limbs. It was moderate in degree in the left arm and more severe in the legs and right arm. The latter is used very little and all movements are weak and show gross inco-ordination. No fine movements of the fingers are possible and any movements of the larger joints tend to result in a gross increase of tone in the limb which inhibits further activity. Finger movements with the left arm are very poor but she was able to grasp small objects after one or two attempts and to hold them and release them. The hands were used with all the fingers in action in the infantile manner.

In both arms there was contracture of the pronator muscles of the forearms which limited both voluntary and passive supination. Other passive movements were full.

In /

In the legs there were contractures at the ankles limiting dorsiflexion of the feet and slight contracture of the right knee in flexion. She kicked the legs quite freely from the hips but movements at the knees were limited and all movements were inco-ordinate and clumsy. When she "walked" with support there was a tendency to equinus at the feet, scissoring at the hips and the feet were placed in front of one another persistently. She could not move the toes voluntarily.

There was an increase of tone in the legs, most marked in the right leg which w as of rigid type and moverately severe. There was a slighter increase of tone in the right arm and an increase of tone less in degree in the left arm, also of rigid type. The biceps, triceps, supinator knee and ankle jerks were increased bilaterally, those in the legs being brisker than those in the upper limbs, and on the right side than the left. The Hoffmann signs were negative bilaterally and the Babinski signs were positive bilaterally.

When the child was standing with support the arms were held in constrained positions, the right in a position of adduction at the shoulder, extension at the elbow, pronation of the forearm and flexion at the wrist and fingers. The position of the left arm was similar but the elbow was semi-flexed so that the hand was across the abdomen. The tonic neck reflexes were positive. When she was held horizontally, the head was thrown backwards, the back arched and the whole body became stiff, the child showing extension of the legs with bilateral plantar flexion of the feet and scissoring, the arms adducted at the shoulder, extended at the elbow, flexed at the wrists and fingers and pronated at the forearm When either arm was moved or when the legs were moved either voluntarily or passively when she lay down, the limbs assumed similar positions to those evident on "walking".

She was re-examined one year after the above examination when she was 4.

At this time the rigidity of all the limbs at rest had increased and the paroxysmal rigidity on any voluntary movement was less marked. When she was held in the walking position, the position of the arms was similar to that previously noted except that the flexion of the left elbow was more marked and there was slight flexion at the right elbow also. The tonic neck reflexes were present but less marked. No extensor thrust of the trunk and limbs could be elicited by holding her horizontally. The movements of the arms noted to occur on voluntary or passive movement of the opposite upper or either lower limbs when she lay prone were still evident, but again there was some flexion at the right elbow and the right /

right stiff limb was not thrust backwards to the same extent as before. Its position resembled more, that of the left arm previously noted.

Sensory findings were untestable but pin prick was appreciated in all areas. The hands and feet were very cold and the feet on one occasion, slightly cyanosed.

Clearly with dystonia of this severity well co-ordinated voluntary activity is, and will remain, impossible. The child is thus very severely handicapped.

Cases in the rigid and spastic stage.

Of the 79 cases classified as diplegias, 72 were in the stage of rigidity or spasticity, with or without contracture, at the time of examination. The numbers of paraplegic, tetraplegic and triplegic cases and their classification into severe, moderately severe and slight categories is shown in Table XII. That the degree of functional impairment is roughly related to the number of limbs involved is shown in Table where the times at which the child first walked and talked is noted.

That the majority of cases of tetraplegic are unable to walk unsupported before the age of 3 indicates how severely these children are handicapped.

As with hemiplegia the reasons for the impairment of function will be considered firstly from the point of view of the actual disturbance of the motor control of the limbs and then from the point of view of the child's inability to use them fully. Other associated abnormalities encountered in patients with diplegia will be discussed.

The impairment of voluntary movement. /

The impairment of voluntary movement.

The causes of the impairment of voluntary movement will be considered under the headings of loss of voluntary power, involuntary movements, rigidity, contracture and other disturbances. It is even more difficult in many cases to assess wo what degree the imppairment, the power and co-ordination of voluntary movements is due to each of the various factors involved, in diplegia than it is in hemiplegia.

Loss of voluntary power.

Loss of voluntary power occurs in even the mildest cases of diplegia and in mild triplegic and tetraplegic cases it is the main cause of impairment of function in the upper limbs.

The loss of voluntary power may be regarded as a result of retardation or failure of the normal development of voluntary motor control. Thus the motor control in cases in the hypotonic stage of diplegia resembles that of the new born child. In the dystonic phase and the early rigid stage, severely affected patients show a development of motor control similar to that found in the normal child to the age of about 8 or 9 months. The hands are beginning to be utilised in conscious activities but thumb and forefinger movements are little used. The onset of the stage of spasticity coincides with the appearance of more highly developed adult types of movement patterns which are impaired by the motor disability.

The impairment of voluntary power is most evident in later developed /

developed movements and in the periphery of the limbs. Movements of the thumb, extension of the fingers, wrist and supination of the forearm are movements that are always weakened and sometimes limited in triplegia and tetraplegia. Voluntary toe movements are almost always absent or grossly impaired in diplegia. In early cases without considerable contracture, it may be possible to demonstrate that dorsiflexion of the foot is limited in extent though no local cause for the limitation is apparent. There is an apparent loss of the movement pattern.

In severe cases of tetraplegic paresis, the impairment of voluntary power is evident, even in movements of quite primitive nature and of the proximal parts of the limbs. Unfortunately from the clinical point of view the loss, or more accurately, failure to develop of actual movement patterns is overshadowed by the impairment of movement which results from later developing rigidity, involuntary movements, rigidity and contracture.

The loss of movement patterns is seen more clearly in cases of diplegia with cerebrellar ataxia where rigidity tends to be less marked and in which very gross loss of the ability to move the fingers, toes, hands and feet voluntarily may be observed.

Associated movements.

The impairment of motor control that results from associated movements is due to three types of movement in diplegia. Movements /

Movements of the dystonic type and athetosis, similar to that found in hemiplegic cases are common. Movements of choreoid type are rare. Movements of dystonic type as a cause of impairment of motor function are, of course, most marked in the dystonic stage of diplegia and have been discussed in the section dealing with this phase of the development of the condition.

Sometimes, however, involuntary movements recognisably of similar pattern to the more extensive dystonic movement of the dystonic stage, persist into the rigid phase and are severe enough to cause severe impairment of voluntary movements. The stimuli of the erect position, pressure on the soles of the feet and the mixture of postural control and voluntary movement found when severely affected children began to walk were sufficient to elicit these involuntary movements, in a number of cases.

In these cases there was a very marked tendency for sudden extension of the neck, back and limbs to occur whenever attempts were made to take a step unsupported. Balance was immediately lost and retardation in achieving unsupported walking balance was often severe, though when the child was lifted so that the stimulus to involuntary postural movements of pressure on the soles of the feet was absent, walking movements of the legs might be perfect.

Case 34. Male. born 1949.

A case of severe tetraplegic rigidity.

The fourth child of mother aged 36 at the time of delivery.

The first child was a forceps delivery, the second born with /

with a labour of less than one hour. Both were born at term. Both healthy. Third child stillborn as a result of concealed/ revealed haemorrhage from placenta previa.

She was well in her fourth pregnancy. She was given ergot tablets at term and an injection of pituitrin. One hour after the latter the child was born precipitously while she was sitting in a chair, as the result of a single pain of great severity. The child weighed 7 lbs.

The child did not breathe for about 20 minutes and did not cry for over one hour and then very feebly. He was blue for almost two days. He had to be tube fed for over one week. Thereafter he was bottle fed but swallowing was very difficult and he often coughed up milk during feeds. At the age of 3 or 4 days he began to show sudden attacks of stiffness in which he arched the back and neck and extended the limbs. "Like a stiff doll, not like a baby at all". The attacks occurred whenever the child was moved, when he choked, and sometimes apparently when he was in his cot alone. The attacks continued until the age of two, by which time he w as able to hold up the head. He succeeded in sitting with support at the age of $3\frac{1}{2}$. At the age of 4 he began to hold toys in the left hand but movements of this were very clumsy and he frequently dropped them.

His limbs were noted to be becoming stiff shortly after the age of one year. The stiffness appeared first in the legs and has remained more marked in them than in the upper limbs, though these are also affected. The stiffness is now marked and at time he "is like a matchstick man with his arms screwed on the wrong way round, and his hands pointing to the back".

On examination at the age of 4 he was $33\frac{1}{2}$" high and head circumference was 18". His teeth were very decayed. He could obey simple commands; trying, unsuccessfully to speak; drooling and swallowing infrequently. Appeared to see and hear normally. Bilateral abducent paresis, more marked on the left than the right. The face was rather expressionless and immobile, but not asymmetrical. Swallowing was slow, infrequent and obviously difficult. Tongue movements in all directions were slow and restricted in range. The grasp of the hands was fair. The whole hands were used and there was no attempt to hold objects with the finger and thumb alone. The thumbs were contracted into the palms though not severely. The forearms showed bilateral pronation contracture. Dorsiflexion of the feet was impossible on account of contracture of the tendon Achilles.

There was generalised rigidity of the limbs, much more marked /

Case 34.Severe tetraplegic diplegia in an early stage of rigidity.He shows extensor positions of all the limbs and is holding the upper limbs anteriorly to prevent them being extended behind the trunk,as is found during dystonic attacks.

marked in the lower limbs than the upper. The tendor jerks were very greatly increased in all the limbs. The knee and ankle jerks were more increased, relatively than were those of the upper limbs. The plantars were bilaterally extensor. Stretch responses were evident in all limbs.

Whenever he attempted voluntary movement of the fingers severe athetosis of the fingers and wrists was evident bilaterally. Whenever movements of the whole upper limb were attempted there was a marked tendency to associated extensor movements. These were severe in type and resulted in the upper limbs being adducted at the shoulders, extended at the elbows and pronated at the forearms. The wrist and fingers were extended. ("Arms on the wrong way round") the legs were extended at the hips, knees and the feet were plantar flexed. Slight scissoring was apparent.

When he was placed on his feet in the upright position, the head was suddenly extended, the back arched, the limbs became markedly extended. All attempts at standing balanced was vitiated by a form of dystonic attack. When he was held off the ground, however, the dystonic position was not in evidence. The legs were placed in a similar position to that found when the feet were placed to the ground. But no extension of the back or neck was evident. The upper limbs were not strongly extended but rather tended to be held in flexed positions.

It was impossible to produce dystonic attacks by extension of the head unless the pressure was exerted on the soles of the feet, either by placing a hand against them, or by placing them on the ground.

Sensory findings could not be tested fully but pin prick appeared to be appreciated in all areas.

Only when the degree of motor control was sufficient to overcome the tendency to neck extension caused by pressure on the soles of the feet, the erect position and massive walking movements, was walking achieved.

The involuntary movements of extension tended to be especially severe in cases in which all four limbs were severely involved /

involved and in these children the use of an upper limb might be grossly restricted because of them. Objects could be grasped only very clumsily and sometimes the extension of the arm was severe and sudden enough to result in it being thrown out of the hand once grasped.

It was interesting to observe that on examination it was possible to produce isolated movements of extension in the limbs of the type described in 4 of the cases classed as paraplegic, 6 of those classified as triplegic and 9 of those classified as tetraplegic, and this in spite of the presence of contracture in the latter cases. The most potent stimuli were found to be extending the head and neck, exerting pressure on the soles of the feet and maintaining the child in the erect position. In all these cases and in all the tetraplegic cases the tonic neck reflex could be relied upon to produce a more or less easily elicited extension in the limbs on the side from which the head was turned.

In a few cases the movements of dystonic type occurred until very late in the rigid and spastic stages of diplegia, and resulted in great functional impairment of voluntary movements.

In most cases, however, the dystonic types of involuntary movement disappeared when spasticity of the limbs became apparent, after a longer or shorter period of time. In many cases, however, they were superceded by athetosis in the spastic limbs leading to almost equally severe derangement of function. This athetosis is identical in features to that described /

described in hemiplegic cases. In some tetraplegic cases it is bilateral, in others one limb may be spared, or may show the more primitive type of extensor associated movement. As in hemiplegia the degree of functional impairment it causes varies greatly.

Case 147. Male. Born 1950.

A case of moderately severe triplegic paresis.

The first child of healthy parents. Mother aged 31 at the time of delivery. She had severe preeclampsia in the last 6 weeks of her pregnancy. She went into labour at term and was admitted to a Nursing Home, but after a few hours pains ceased and she was given pituitrin injections to restart them. These injections were unsuccessful, however, and she had an artificial rupture of membranes three days later, after which labour pains commenced. The child was delivered by mid cavity forceps delivery.

The child weighed 6 lbs. 12 ozs. He was born crying and seemed well until the seventh day when he had a generalised convulsion. Twitching of the limbs continued for four days. Thereafter he seemed to improve and was discharged from the Nursing Home at the age of 4 weeks.

Apart from crying a great deal he appeared to be normal. At the age of 6 weeks, however, he sometimes seemed stiff in the mother's arms. This was most evident when he was being bathed and the head was held back. Mother does not remember position of the limbs.

At the age of three months he was able to hold up the head. By this time the legs were definitely stiff and the right hand seemed to be stiff, the fingers clenched and the limbs moved less freely than the left. The child could sit without support at the age of 15 months and shortly after began to propel himself along with the left arm, while in the sitting position. He was able to walk with support at the age of 3. He had begun to speak at the age of 9 or 10 months.

On examination at the age of $3\frac{1}{2}$ he was an intelligent spoilt boy with spastic triplegia.

Height was $34\frac{1}{2}$". Head circumference was 20".

His speech was normal except for slight slurring of consonants. /

consonants. Its content was good. There was bilateral abducent weakness. Slight right facial weakness was evident on movement only. The tongue deviated to the right on central protrusion.

There was impairment of power in all the limbs. This was moderately severe in the legs and right arm and much less severe in the left upper limb. Full abduction of the thumb and supination of the forearm was limited on the right and the latter slightly limited on the left. Dorsiflexion of the feet was rather severely limited bilaterally. Movements of the left hand were rather clumsy but probably not outwith the range of normality. In the left hand, however, all movements were accompanied by a marked degree of athetosis of the fingers which were severely hyperextended. The wrist showed some involuntary extensor movement at the same time as the fingers. The involuntary movements of the fingers and wrist were slow and writhing in type. He was able to grasp with the left hand when made to, but was unable to release the grasp.

There was a marked spastic increase of tone in the left arm and in the lower limbs, the increase in the latter being more severe. There was a slight spastic increase of tone in the right arm. The biceps, triceps and supinator jerks were much increased in the left arm and slightly in the right. The knee and ankle jerks were much increased bilaterally. The plantar responses were extensor.

Sensory findings. Could not be tested fully, but no impairment was demonstrated.

The choreoid type of associated movement, which occurs occasionally in hemiplegia also occurs, but rarely in cases of diplegia in the spastic stage.

The movement is more rapid, and generally more localised than the more primitive dystonic form and it is much more frequent in the upper limbs than in the lower. Two cases only showed choreoid associated movements in the present series and in one they were very slight.

Rigidity of the limbs. /

Rigidity of the limbs.

A cause of limitation of voluntary movement not found in cases of hemiplegia is generalised increase of tone not due to contracture. This is almost invariable in cases of diplegia of moderately severe or severe degree. Indeed the earlier authors doubted if there was in fact any real loss of movement patterns in diplegia, and attributed the limitation of voluntary movement only to rigidity. (Little, 1862; Freud, 1893).

The fact remains that rigidity is a very important cause of loss of range and power of voluntary movement, in paraplegic, tripligic and tetraplegic cases. The rigidity of the limbs appears before the dystonic attacks have ceased, or in paraplegic cases with normal milestones about the age of 3-4 months. It is usually first evident in the thighs and knees, but rapidly spreads and increases in the lower limbs, and involves the upper limbs also. Initially marked variations from day to day and minute to minute may be found. When the child is escited, especially when feeding, the rigidity may be very pronounced. When the child is held in positions which are liable to produce dystonia in susceptible children the rigidity is accentuated. The most potent means of accentuating rigidity has been found to be to extend the head. The easiest way to produce diminution in rigidity has been found to flex the head strongly. This latter manœuvre has been much utilised by some schools for producing muscular relaxation in diplegic children. (Bobarth and Bobarth, 1950).

In /

In the same way as the assumption of the upright position, pressure on the soles of the feet and attempting voluntary movement produces associated extension positions in the limbs so they also tend to produce generalised increase of tone in the muscles of the affected limbs, whether or not, in fact dystonia has been a feature in the individual patient.

The rigidity is always much more pronounced in the lower limbs than the upper, whether the case is slightly or severely affected. In moderately severe and severe cases it may be enough to inhibit all attempts voluntarily to move the limb.

The reflex nature of the rigidity is shown by the fact that voluntary movements may be rendered quite impotent by its presence in some positions of the child, whereas movements may be quite full in range if the child is placed in positions where his rigidity is diminished. The following case had the greatest difficulty in walking at all because of the extremely severe rigidity of his legs. Yet when placed on his back with the head strongly flexed the range of leg movements was good and their power only moderately impaired. The rigidity had been much reduced by the manœuvres employed.

Case 25. Male. Born

Severe spastic tetraplegia.

The first of 4 children born to mother aged 19 at the time of delivery and her healthy husband. Three subsequent children born after normal pregnancy and delivery are well. Mother and the maternal aunts and grandmother are left handed.

Mother was well during pregnancy until 4 weeks before the expected /

expected date of delivery when she had a severe vaginal haemorrhage. She was admitted to hospital where a placenta previa was found and Willetts forceps were applied to the foetal head. Delivery occurred after a labour of 2 hours and was accomplished under anaesthesia. The placenta showed numerous infarcts and weighed 1 lb. 5 ozs. The child was limp following delivery but responded to oxygen, though on admission to the premature nursery was said to be "rather murmury". The birth weight was 5 lbs. 15 ozs. After 2 days in the premature nursery the child appeared to be normal and breast feeding was begun. He was discharged on the 13th. day.

He was said to be a good baby. It was noted that the left eye was turned in. He sat with support at the age of 8 months and alone shortly afterwards. He began to crawl at the at the age of 2 and when he did so it was found that he pulled himself along with the arms, pulling his legs behind him stiffly "like a tail". He was able to stand at the age of 3½ and to walk without support at the age of 4½. When he began to walk he was very unsteady and fell a great deal because he kept putting one foot in front of the other and tripping. Gradually his gait improved and by the age of 5 he was able to walk without falling more than occasionally, though he still proceeded on the toes, not putting the heels to the ground. With lengthening of the tendon Achilles operatively at the age of 5½ the gait improved further and he manages to get the heels to the ground.

Mother thinks he was rather slow to use the hands and used the right hand before the left. He always handled objects poorly and at the age of 11 could still not do all his buttons or his laces. His right hand was better than his left.

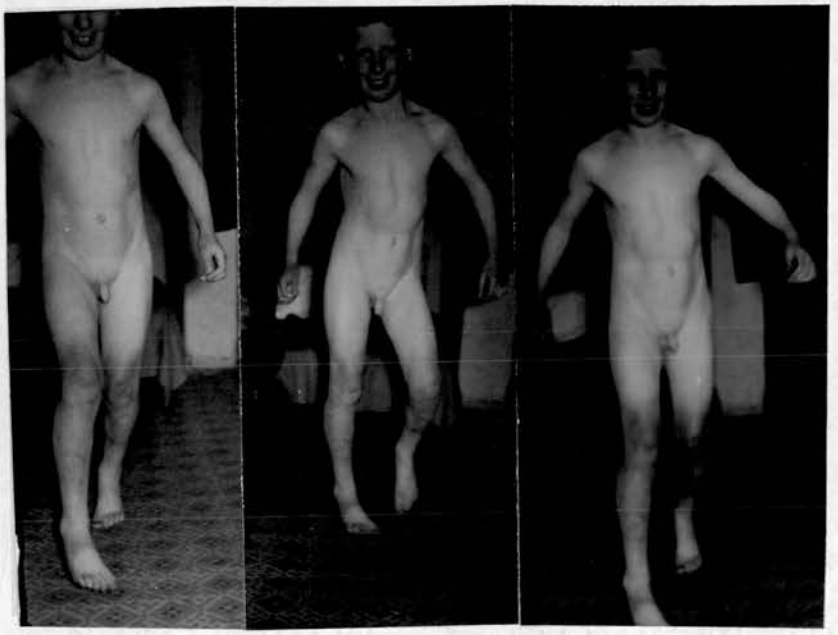
He said his first words at the age of 18 months and his speech developed normally thereafter.

At the age of 6 he went to a school for the physically handicapped and has done moderately well there, learning to read and write without apparent difficulty. I.Q. 82.

Of quiet, placid disposition, but able to play successfully with children of his own age.

Examination. Height 55½". Head circumference 21½".

He was slow in response to commands and in answer to questions but accurate. His comprehension was good but his speech, though of fair content, was rather slurred and jerkily /



Case 25.The gait in a case of tetraplegic paresis.The rigidity of the limbs is so severe that accurate voluntary movements are impossible in the legs.The child is unsteady and there is much trunk movement in an attempt to compensate for the poor limb movements and to maintain balance.

jerkily pronounced.

There was a slight right facial weakness shown by lag on voluntary and emotional movement though the face was symmetrical at rest.

The power and co-ordination of the limbs was impaired. The impairment was more marked in the legs than the arms. There was limitation of passive supination of the left forearm, but other passive and voluntary movements were full though supination of the forearm, extension of the fingers, wrists and elbows and abduction of the shoulders were weak bilaterally, though more impaired on the left than the right.

Fine and rapid finger movements were poor bilaterally and though he could handle small objects he used the whole hand in an infantile way without especial use of the thumbs and forefingers. The finger movements are better controlled on the right than the left.

There were flexor contractures of the hips, knees and ankles preventing full hip and knee extension and full dorsiflexion at the ankles. The contractures were more severe on the left than the right. The gait was spastic with scissoring of the legs, flexion and adduction of the hips, flexion of the knees and a tendency to equinus position of the feet.

As he walked he leant forwards and the arms were in perpetual motion tending to be held adducted at the shoulder, flexed at the wrists and fingers, pronated at the forearms and flexed to variable degrees at the elbows. He had great difficulty in maintaining his balance when he initiated his first step.

There was a marked increase of tone in the limbs. This was moderate in the right arm, rather more severe in the left arm and very severe in both legs. The tone was spastic with underlying rigidity. Stretch responses were present in all limbs. The biceps, triceps supinator jerks were moderately increased, more on the left than the right and the knee and ankle jerks were much increased with clonus at both knees and both ankles.

The Hoffmann signs were present bilaterally. The plantar responses were extensor on right and left. The fingers showed some hyperextensibility bilaterally.

The tonic neck reflexes were present and well marked. When he used either upper limb forcefully the contralateral arm /

arm showed a tendency to assume a position of adduction at the shoulder, flexion at the elbow, wrist and fingers, and pronation at the forearm.

Sensory findings were entirely normal. The feet and hands were extremely cold and tended to become slightly cyanosed.

Though much less severe in degree in the upper limbs it is severe enough to limit the power and extent of voluntary movements in a large proportion of cases. Again the rigidity may be reduced by head flexion, a fact soon learnt by many severely affected children who are often mistakenly thought to have visual defect because of their strongly flexed heads at their desks at school.

The rigidity is more marked in the proximal than the distal parts of the limbs in most cases, and in many the hands and feet show little increase of tone until the period of contracture involves them during the spastic stage of diplegia.

Unfortunately when spasticity occurs it tends to be superimposed on the rigidity rather than to replace it. It is difficult to be sure to what degree the rigidity recedes in time. Certainly the changes in its severity in different positions of the child becomes less marked, or they are masked by the spasticity. The presence of the rigidity and spasticity together in cases showing flexor associated movements, increase in tendon jerks and extensor plantar responses certainly accounts for the characteristic feel of the lower limbs if diplegic patients even in the late stages of their disorder.

The /

TABLE XIV

The ages at which 79 patients with diplegia walked without support.

Distribution of paresis	Tetraplegia	Triplegia	Paraplegia	All cases
Age				
Under 18 months	0	2	7	9
Over 18 months under 3 yrs.	4	6	15	25
Over 3 years and under 6 yrs.	9	8	3	20
Over 6 years and under 8 yrs.	3	1	0	4
Over 8 years	2	1	0	3
Not walking	5	3	3	11
Unknown	4	2	1	7
	27	23	29	79

TABLE XV

The age at which 79 patients of dyplegia said their first words.

	Distribution of paresis			
	Tetrapegia	Triplegia	Paraplegia	All cases
Age				
Under 18 mths.	3	9	18	30
Over 18 mths. under 3 yrs.	9	7	7	23
Over 3 yrs. Under 6 yrs.	3	4	3	10
Over 6 yrs. Under 8 yrs.	2	0	0	2
Over 8 yrs.	0	0	0	0
Not walking	8	1	0	9
Unknown	2	2	1	5
	27	23	29	79

The spastic phase of diplegia and the importance of contracture in limiting voluntary movements.

The alterations that occur with the onset of spasticity in cases of diplegia are most fascinating clinically. The stage of the disorder at which spasticity becomes evident usually coincides with the child's first attempts to use the thumb and forefinger, and with his ability to control the postures of his limbs so as to allow more efficient handling of objects to be possible.

At the same time voluntary movements cease to result in the associated movements of pure extension. Some flexion is first evident in the elbows and over a period of weeks or months the flexion spreads and becomes more marked so that the whole of the involved upper limbs assume positions similar to those found in hemiplegic patients, on voluntary movements.

It is difficult to see that the appearance of spasticity itself has any effect on the functional use of the involved upper limbs. The lack of power and range of movement is evident before spasticity appears. In the lower limbs the greater difficulty in moving from the hips does seem to have some some deleterious effect in some cases, and in two the histories suggest that children beginning to walk temporally retrogressed at this stage.

The great importance of the appearance of spasticity from the functional point of view is the rapidity with which it may be complicated by severe contracture in the limbs. The same factors as favour the appearance of spasticity in hemiplegia also favour it in cases of diplegia. Since the

history /

history in diplegic cases tends to be less dramatic than in many cases of hemiplegia these cases tend to remain without adequate medical treatment for longer. In fact, though the majority of cases of diplegia were taken to their doctors very shortly after the appearance of their first symptoms there was an average delay in their receiving adequate medical treatment of almost two years. In almost all cases contractures of some severity were present by this time.

As in hemiplegia the contractures tend to be most marked in the periphery of the limbs than the proximal. This leads to a very interesting phenomenon in diplegia of hands, clumsy, but otherwise apparently unaffected becoming relatively useless in quite a short space of time. Whereas before the onset of contracture the child usually finds abduction and extension of the thumb and fingers difficult when contracture ensues, these movements may become impossible in a matter of a few weeks.

Pronation contracture of the forearms in those with upper limb involvement is invariable in those with much spasticity, the elbows and shoulders are less affected. In contrast to hemiplegia the legs are more involved than the arms. The feet become fixed in a position of plantar flexion, the legs and hips in flexion, or variable degrees of severity.

The age of the appearance of contractures is extremely variable. As has been noted, the spastic phase may be delayed /

delayed in its appearance for up to 4 years, and probably longer. Some contracture has been noted as early as six months, but this is unusual. Very severe contractures may be present by the age of two years.

Typical positions of contracture are shown by the following case.

Case 55. Male. Born 1939.

Rigid spastic diplegia with cortical blindness and severe mental defect.

The second of three children born a mother aged 34 at the time of delivery.

The first and third born children were born after normal pregnancy and are well. Both labours were protracted on account of rigidity of the maternal soft parts.

In her second pregnancy she was constantly severely worried on account of the failure of her husbands business and the fact that he was due to be called up.

The delivery occurred at term and followed a rapid, severe labour of about 10 minutes at home. The child weighed 7 lbs. 7 ozs. and the placenta weighed 1 lb. and showed no infarcts. The child cried at once and seemed normal. He was very slow in feeding and seemed reluctant to swallow milk once in his mouth. He slept most of the day and night. At the age of 6 weeks he began to have attacks. In these he suddenly became stiff with the head and trunk thrown backwards, the legs extended and the arms flexed, and then suddenly flexed forwards violently and became limp, with impairment of consciousness for a few seconds. The attacks seemed to be precipitated by handling, but occurred inconstantly from to 20 times a day. When he had a cold or other infection they became more frequent. The attacks persisted, similar in their original form until the age of about 4 years.

The child never looked about him. He held up the head at the age of about 18 months but never achieved a sitting position, never comprehended anything that was said to him and never spoke. Weaning proved impossible as he could not swallow solids. Photographs taken at the age of 2 showed a small square headed boy being held up by the shoulder, the legs extended and the feet crossed, the right arm adducted at the /

the shoulder, extended at the elbow, pronated at the forearm and the wrist and hand flexed, the left arm flexed at the elbow but otherwise in a similar position to the right.

He was admitted to an institution for mental defectives at the age of 4 and his vision over the years gradually improved. He was able to recognise people at the age of 8 and swallow solids about the same age. Unable to sit even with support.

Examination. Length 48½". Head circumference 19½". The head showed gross occipital flattening. The limbs were equal in length. High palate. Scattered milia over head and trunk.

He was able to recognise his nurses and was worried by changes of place. He apparently comprehended no speech and had none, except that he recognised his name. He was bedridden by contractures. He had some vision, recognised light and dark and could perceive the movements of large masses. The fundi showed very white discs but no other abnormality. The face was rather expressionless and immobile but symmetrical.

He swallowed with an effort, infrequently and showed much drooling.

The power of all the limbs was grossly impaired. The impairment was more marked in the lower limbs than the upper. The range of purposive movement was negligible. There were flexion contractures in the hips and knees bilaterally and the feet were fixed in plantar flexion. The fingers were placed over the thumb, pressing into the palm of the right hand and flexion contractures of the right elbow and wrist were present. He could be persuaded to grasp a little with both hands but the grasp was uncertain and he had difficulty in releasing it. The movements of the legs were weak and restricted by contracture bilaterally.

There was a very marked mixed spastic and rigid increase of tone in the legs and less marked in the right arm. In the left arm the increase was predominantly rigid, and less marked.

The biceps, triceps and supinator jerks were somewhat exaggerated on the left and very marked on the right. In the legs and knee and ankle jerks were very brisk indeed and bilateral ankle clonus, rather difficult to elicit, was present. The Hoffmann signs were absent. The movement of the great toe was extensor to practically every stimulus, including Oppenheim and Chaddock tests, but flexor on plantar stimulation.

When /

Case 77. Severe tetraplegic contractures in a diplegia girl aged four and a half years.

When held upright the legs crossed and assumed a position of extension, in so far as the contractures would allow, with the feet crossed. The right arm showed adduction at the shoulder, flexion at the elbow, wrist and fingers, and pronation of the forearm. The left arm showed a similar position except that there was a marked extension at the elbow. The tonic neck reflexes were bilaterally present, more marked in the left arm. A slight extensor thrust, with the head thrown back and the trunk in a position of opithotonos, the arms being extended at the elbows could be elicited. The hands were soft and the fingers were grossly malleable and hyper-extensible.

Sensory findings untestable.

From the point of view of function, however, the most severe results of contracture are often found in the hands, where movements already weak and poorly co-ordinate are further grossly impaired.

Of the 79 cases of diplegia in the series, 28 had already had tendon lengthening operations by the time of examination, others were obviously going to require them. Yet it was apparent that in a number of cases the necessity for operation had been avoided by early diagnosis and physiotherapy though their paresis was as severe as in the former group.

In the legs it was often extremely difficult to assess to what degree the stiffness of the legs was due to true muscular rigidity of reflex origin and how much to contracture. This was especially true of the marked plantar flexion of the feet that was found in severe cases of diplegia. The degree of flexion contracture evident at the hips and knees was not a reliable guide as it depended so much on the degree of disuse /

Case I67. Patient with severe diplegia in the stage of spasticity.There are contractures in all the limbs of flexion type.He was in an institution for the mentally defective.

disuse to which the limbs had been subjected.

Contracture was more marked, a s was to be expected in older patients than in younger. As in hemiplegia, contracture appeared to be somewhat exacerbated about the time of puberty. (Féré 1896).

Dwarfing in diplegia.

The marked dwarfing of the pelvis and lower limbs evident in diplegic patients was noted by the early observers of the condition. (Little, 1843; 1862; Gowers, 1888; Freud 1893).

Its severity roughly parallels the severity and extent of the paresis of the limbs, and was more marked in tetraplegic cases than in paraplegic. Unfortunately comparable figures for male, female, premature and full time infants at various ages are not yet available for normal Edinburgh children.

In view of the fact that the most severely affected limbs show marked retardation of growth in diplegia, it might be expected that in severe cases of triplegia, dwarfing of the affected upper limb as well as of the lower would be apparent. In fact this was not found. Only one case of triplegia showed apparent shortening of more than $\frac{1}{2}$" in the more affected upper limbs compared to the less affected. This is in accordance with the observations of Freud (1897).

Why dwarfing of the affected upper limb should occur in hemiplegia and not in triplegia is difficult to see. It is the more difficult to account for it when the similarity of

some /

some of the other findings in hemiplegia and triplegia are considered. Many of the latter, in fact, have a superficial appearance of paraplegia with superimposed hemiplegia. Spasticity of the upper limb rather than rigidity is evident; the tendon jerks are much increased; the Hoffmann response is frequently positive; athetosis may be present; the associated movements are flexor in type; the contracture is similar to that found in hemiplegia. Yet dwarfing in the more affected upper limb is exceptional in triplegia and usual in hemiplegia.

Case 103. Female. Born 1939.

Moderately severe spastic triplegia.

The only child of an unmarried mother aged 26 at the time of delivery.

Apart from being very worried the mother's health was normal during pregnancy. The child was born at term after a labour of about 24 hours spontaneously. She was blue at birth and breathed only after resuscitation had been employed for about 15 minutes. The child was restless and showed twitching for 7 days. Thereafter her state appeared to be normal and she was discharged from the Maternity Hospital at the age of 3 weeks.

She was a quiet baby at home. Fed well and slept but was rather inactive.

She sat without support at the age of 14 months and about this time the mother, wondering why the child could not walk, noted that the legs were stiff, though the hands were used fairly well. She was able to walk with support at the age of 5 and without it between the age of 6 and 7. She said her first words at the age of 2 but could not form sentences before the age of 7.

Attended school for the mentally defectives from the age of 7. Poor progress. I.Q. 54.

Attended clinic for the mentally defective from the age of 18 months where sudden attacks of hyperextension of the back with extensor positions of the limbs occurring many times a day with changes of position were noted until the age of 3.

Examination /

Examination at the age of 13½. A rather dull pubescent child. Height 61". Weight 84 pounds. Head circumference 19¾". No of skull, trunk or limbs. Speech of very poor content. Replies to questions and commands slow and inaccurate. Speech defective, multiple defects of consonant pronunciation. Vision and hearing normal.

Bilateral abducent paresis. Slight left facial paresis of upper motor neurone type only evident at rest. Movements of all limbs very clumsy and weak. On the left more definite weakness and inco-ordination of fine movements especially could be demonstrated. Supination of the left arm was limited and very weak, that of the right normal. Extension of the left fingers slightly impaired and very weak. Full extension of the left arm at the elbow voluntarily was impossible.

Whenever she walked or used the right arm, the left immediately assumed a position of flexion at the elbow, wrist and fingers, pronation of the forearm and adduction at the shoulder.

There was a moderately increased tone of the left arm and both legs of spastic type. This was not evident in the right arm; it was more marked in the legs than the arm.

The biceps, triceps and supinator jerks were increased on the left compared to the right. The knee and ankle jerks were very brisk bilaterally. The Hoffmann response was positive on the left and negative on the right. The plantars were bilaterally extensor.

Sensory findings. She appeared to appreciate pin prick in all areas and no pass pointing could be demonstrated. No temperature difference was noted in the limbs.

It is interesting, however, to note that in triplegic patients it is also exceptional for there to be significant temperature difference between the hands.

In the majority of cases of diplegia the head was smaller than was to be expected on the basis of current tables of head size. Since over one third of the patients were prematurely born, however, more detailed study of the results of head measurements /

measurements must await the availability of comparable Edinburgh figures. Seven cases, four tetraplegic, two triplegic and one paraplegic, showed a difference of ½" or more in the cranial hemicircumference. A much larger number showed quite marked plagiocephaly, which most disappointingly, gave very similar measurements for each hemicircumference.

Other neurological findings in the limbs in diplegia.

Vaso-motor changes in the limbs in diplegia were sometimes evident in the feet but rarely in the hands and in no cases was marked difference between the sides noted. In most cases the feet felt colder than the hands, and pallor was sometimes evident but the findings were not striking.

In one case only was definite sensory impairment demonstrated, a loss of joint sense in the lower limbs. Very slight loss of joint sense w as almost universal in those whose limbs were immobile but in no case receiving routine physiotherapy was even this discovered.

Neglect of the more severely affected limb, similar to that found in hemiplegia was not found to be a feature of diplegia though in a few cases detailed questioning of the mother gave the impression that neglect of the more severely affected side might be present transiently following a fit, especially in triplegic patients.

Mental impairment in diplegia.

As in hemiplegia the estimation of intelligence by tests in /

TABLE XVI

The intelligence quotient in 79 children with diplegia.

	No. of cases	Intelligence quotient						
		115+	100–114	85–99	70–84	55–69	Under 55	Untested
Paraplegia	29	1	5	4	6	7	1	5
Triplegia	23	2	2	0	5	3	3	8
Tetraplegia	27	0	0	2	4	3	11	7
All cases	79	3	7	6	15	13	15	20

TABLE XVII

Schooling in 79 patients with diplegia in childhood.

	No. of cases	Normal schools	Schools for the physically handicapped.	Schools for the mentally handicapped.	Ineducable	Not yet at school
Paraplegia	29	7	9	5	0	8
Triplegia	23	3	3	5	4	8
Tetraplegia	27	1	0	6	9	11
All cases	79	11	12	16	13	27

in diplegic children is fraught with uncertainties. The success of the child in achieving social adaptation and educational standards is probably equally fallible as a guide to intellectual impairment. (Dunsdon, 1952).

The schooling received by diplegic children and their intelligence as estimated by different psychologists and tests in various circumstances are shown in Tables XVI & XVII It must be emphasised that the test results are not to be taken as accurate representations of the actual intelligence of all cases because the method testing varied so greatly. Moreover 20 patients, 4 of whom were in normal schools were not tested for various reasons. The most that can be taken from the results in the Table is that the numbers of children with severely impaired intelligence is greater in tetraplegic cases than in paraplegic, and that a higher proportion of the more severely paretic cases are ineducable, than of the less severely affected. The small number of children with diplegia attending normal schools is striking.

The severity of intellectual impairment as measured by intelligence testing was not always a true reflection of a child's inability to achieve some success in the social sphere. And the achievement of some institutional cases was surprising, when the low measurements of intelligence obtained on testing were considered.

To discuss the varieties of mental defect encountered would require much more space and involved description than can /

TABLE XVIII

Speech defects in 79 patients with diplegia.

	No. of cases.	Stutter or hesitation	Dyslalia			No more than one or two words.	Total speech defects.	
			mild	moderately severe	Severe			
Paraplegia	29	0	3	1	2	1	7	
Triplegia	23	4 x	4 x	3	4 x	1	14	
Tetraplegia	29	0	2	4	5	3	14	
Diplegia	79	4	9	8	11	5	35	
x Children showing hesitation or stutter with dyslalia.								

TABLE XIX

Impairment of vision in 79 cases of diplegia.

	No. of cases	Retrolental fibroplasia	optic atrophy	cortical blindness	Total
Paraplegia	29	1	1	0	2
Triplegia	23	3	0	0	3
Tetraplegia	27	2	2 +	4 +	8
Total diplegia patients	79	6 Ø	3 +	4 +	13

Ø/3 with some useful vision.
+ 1 in each category so marked had some useful vision.

be given to it in this essay.

The remarks made about the value of milestones in assessing the degree of mental impairment hold true for diplegia. The most reliable single criterion of mental impairment judged from milestones was delay in talking, and in a number of children with quite severe retardation of talking only slight impairment of intelligence was found, so that this is not a constantly reliable measure by any means. All other functions, however, did tend to be affected more severely and directly by motor involvement, as has already been described. Tables XIV & XV.

Aphasia.

Aphasia was recognised in only four cases of diplegia. Three of these were triplegic, two with predominant upper limb involvement on the right and one on the left. One case was tetraplegic. In no case was the aphasia severe.

A much larger number of mental defective cases of diplegia appeared to have either greater difficulty in comprehension than expression or vice versa. It is probable that a number of these cases did in fact have disturbances similar in type to those classified as true aphasic disorders, but that they were much slighter in degree.

Speech defects.

Thirty five of the children with diplegia showed speech defects of various types. The numbers occurring in paraplegic, triplegic and tetraplegic disorder are shown in Table XVIII It will be seen that speech defects appeared to occur more

frequently in those with tri and tetraplegic disorder than in those with paraplegia.

For the purposes of classification, stutters and hesitations in speech are grouped together. They occurred relatively infrequently. On the other hand, dyslalia, usually abnormal pronunciation of consonants occurred in a large proportion of cases. Mild cases were those in which it was difficult to comprehend speech but not impossible. Moderately severe cases were those in which speech was more or less incomprehensible to those outside the family circle and severe cases those in which speech was more or less incomprehensible to all.

Five cases, all severe cases of paresis, speech was very grossly defective so that the child was incapable of expressing even the simplest ideas. None of these cases was under the age of 5.

As has been noted, the development of speech was retarded to some extent in a very large proportion of those with diplegia.

The importance of speech defects in the education of children with cerebral palsy is accentuated by the handicaps to other means of expression which so many of these children show. (Phelps, 1940; Dunsdon, 1952).

Cranial Nerve Involvement.

The proportion of diplegic patients showing cranial nerve abnormalities with the exception of strabismus, is stated to be /

be low in most surveys. (Osler, 1889; Lovett, 1888). But some early work tended to confirm the present findings that cranial nerve abnormalities, are not, in fact common. (Koenig, 1896).

Cranial Nerves.

Visual defects. Complete or partial blindness was ascertained in 13 cases. In 6 cases retrolental fibroplasia was present. In 3 optic atrophy was present and in 4 field defects were evident, presumably of cortical origin. (Table XIX) (Ingram and Kerr, 1954).

All the cases of retrolental fibroplasia were cases of severe paresis, one paraplegic, three triplegic and two tetraplegic in extent. All the children were born prematurely and all had a history of neonatal anoxia and oxygen therapy.

Optic atrophy with typical retinal appearances, was observed in three cases, one paraplegic and two tetraplegic in distribution. Two of the three cases were blind except for appreciation of light. One had a degree of useful vision, but was severely mentally defective. Three had some useful vision.

Field defects were present in 4 cases. One case showed a complete homonymous hemanopia and had useful vision. The other three cases had vision in only one quadrant and the degree of vision, or the child's utilisation of it was so poor as to be useless.

Of the thirteen cases with blindness, partial or complete, 5 cases /

TABLE XX

Strabismus in 79 diplegic patients.

	Number of cases	Oculomotor weakness		Abducent weakness		Total number of patients
		Unilateral	Bilateral	Unilateral	Bilateral	
Paraplegia	29	1	0	4	4	9
Triplegia	23	0	1	4	6	11
Tetraplegia	27	3 x	3	3 x	6	14
Diplegia	79	4 x	4	11 x	16	34

x one case showed associated unilateral oculomotor and bilateral abducent pareses.

Case 83. Diplegia with tetraplegic involvement of the limbs.

Severe bilateral abducent paresis.

5 cases were considered to show a degree of useful vision, and 8 to have no useful vision.

One case of tetraplegia showed the typical retinal changes of toxoplasmosis, but visual acuity and fields were normal at the time of examination.

A number of cases showed refractive errors of various degrees of severity. Three cases were severely myopic.

Strabismus. This indicates a frequency of strabismus in diplegia in agreement with that noted by early authors. (Heinig, 1896; Freud, 1893). Thirty four patients showed strabismus. The commonest type noted was bilateral abducent paresis, which was very severe in some cases and slight in others. Strabismus was commoner in tetraplegic paresis than in triplegic of paraplegic paresis (Table XX). It was commoner in those born prematurely than in those born at term. Since the strabismus tended to improve as the child grew older, in most cases it was more frequent in its occurrence and more severe in young patients.

In three cases there was bilateral abducent paresis of severe degree which appeared to be of central origin. The eyes could be made to abduct fully involuntarily, but only very slightly on voluntary eye movement.

In another four cases of severe symmetrical bilateral abducent paresis, supranuclear paresis was suspected, but because of poor co-operation of the young children, could not be confirmed on full examination. It is possible that supranuclear /

supranuclear paresis is commoner in diplegia than is generally thought, but the difficulties of confirming its presence in young children are considerable. One case of supranuclear third nerve paresis with inability to close the eyes voluntarily, or adduct the eyes voluntarily, has been observed, but the child was not resident in Edinburgh and is, therefore, not included in the survey. The patient was a girl with severe tetraplegia. One patient with bilateral third nerve paresis in the survey was suspected of having a degree of supranuclear paresis. His eye movements were conjugate on involuntary movement, but not conjugate on voluntary movement and voluntary eye closure was very weak. He was a case of severe tetraplegia.

All the cases with retrolental fibroplasia had unilateral or bilateral sixth nerve paresis of neuromuscular type. Strabismus was evident in all but one of those with partial or complete blindness.

Facial Paresis.

In contrast to some of the earlier statements that facial paresis was unusual in diplegia, facial paresis was found in quite a high proportion of cases of diplegia. This is in agreement with the observations of Koenig,(1896) and Freud, (1893, 1897).

In the series, thirty patients were found to show unilateral facial paresis and three, all cases of tetraplegia, bilateral facial paresis. In only five cases was facial asymmetry /

asymmetry present at rest. In the remaining cases of unilateral paresis the face became asymmetrical only on movement. As with facial paresis in hemiplegia a number of these patients showed overaction of the affected side on movements of expression and underaction of the face on voluntary movements.

In the ten triplegic cases and 13 tetraplegic who showed unilateral facial paresis this was on the side of the most affected arm in all but one case. In those showing bilateral facial paresis it was usual for the paresis to be asymmetrical. In these cases the face tended to be somewhat mask like and moved little on either voluntary or emotional movement.

Swallowing.

A history of drooling and apparent difficulty in swallowing during infancy was elicited in many of the patients with tetraplegia and triplegia.

It was difficult to distinguish between those cases in which the difficulty of swallowing was due to feeding mismanagement and which to organic disorder. The figures ~~in Table~~ refer, therefore, only to those cases in which difficulty in swallowing was evident at the time of examination. In these cases drooling was a prominent feature. Seven of the nine cases who showed difficulty in swallowing were tetraplegic, seven were over the age of three and five were severely mentally defective. All but one showed concomitant facial paresis, either unilateral or bilateral and three showed evidence /

TABLE XXI

Epilepsy in 79 diplegic patients

	No. of cases	Type of attack: Grand mal	Jacksonian	Petit mal	Other	No. of patients
Paraplegia	29	6	0	1	0	6
Triplegia	23	8	1	3	0	8
Tetraplegia	27	6	1	2	3	7
Diplegia	79	20	2	6	3	21 x

x 8 had attacks of more than one type.

evidence of upper motor neurone paresis of the tongue. All those with difficulty in swallowing had speech defects.

In some cases the difficulty in swallowing was so severe that weaning had proved impossible for considerable periods. In some cases swallowing was only evident once in three minutes or more.

The tongue.

The tongue showed defective movement in seven cases, three with triplegia and four with tetraplegia. In three cases the movements appeared to be limited and weak in all directions. In four cases the weakness was asymmetrical. Three of the cases showed difficulty in swallowing and all had facial paresis. All but one of the cases with involvement of the tongue showed defective speech.

Other cranial nerve disturbances.

One case showed impaired ability to elevate one shoulder. She was a case of paraplegia with mental defect, epilepsy and a family history of neurofibromatosis.

Epilepsy.

Epilepsy is a frequent manifestation of diplegia. Many different forms of epilepsy occur and the form of the attacks differs greatly in different cases.

The types which occurred and the frequency of their occurrence in the different forms of diplegia are shown in Table XXI. It will be observed that 21 cases had a history of /

TABLE XXII

The age at the time of the first fit and the last fit in grand mal and petit mal occurring in 79 diplegia patients

		Under 1 year	Over 1 year and under 3	Over 3 years and under 6	Over 6	Present at time of examination
Grand mal	First fit	7	5	6	2	-
	Last fit	0	1	3	0	16
Petit mal	First fit	2	0	2	2	-
	Last fit	0	0	1	0	5

of epileptic attacks. 12 of these had grand mal only, 6 grand mal and petit mal, two Jacksonian attacks and three attacks of different type. In two of the latter grand mal also occurred.

Grand mal alone or in combination with other forms of epilepsy was present in 20 cases. The time of the occurrence of the first fit, the time of the occurrence of the last fit and the frequency of attacks are shown in Tables XXII and XXIII It will be seen that most cases showed epilepsy first before the age of three and that it continued in the majority. All the children in whom epileptic attacks ceased to occur had fits more frequently than once a month during the time it had been evident. None of them showed petit mal.

The severity of grand mal attacks did not seem to have any relationship to the frequency of their occurrence in the small number of cases in the series.

Jacksonian attacks were shown by two cases, one triplegic and one tetraplegic. In each case they began in the upper limb of the more affected side. These patients were extremely interesting in that they tended to show more marked facial paresis on the affected side and the limbs on the side were less useful following attacks. Immediately after attacks the clinical picture was very similar to that of hemiplegic paresis, with neglect of the affected limbs, and an accentuation of the spastic signs on the affected side, except that the /

TABLE XXIII

The frequency of attacks of grand mal in 79 cases of diplegia

More than one attack in a week	2
(Less than one attack in a week (More than one attack in a month	10
Less than one attack in a month More than one attack in a year	6
Less than one attack in a year	2
No. of patients with grand mal.	20

the contralateral lower limbs was also involved more or less severely.

Petit mal occurred in six patients, all of whom showed grand mal in addition. The attacks were present at least once daily in all these cases, and in two occurred more frequently than six times a day.

The cases classified as having attacks dissimilar to those previously described were all cases of moderately severe or severe tetraplegia. In all the attacks were initially associated with dystonic attacks. In one the attacks persisted after dystonic attacks had ceased to occur, in the other two the attacks ceased with the attacks of dystonia. In the latter two cases the attacks began with dystonic movements, usually on handling, similar to those which have been described earlier. The child became stiff, the back, head and limbs extended; but not only was consciousness impaired slightly, but it appeared to be very severely impaired and the child immediately became limp and remained so for a few seconds, before returning to normal. In the third case the attacks began similarly, but the child seemed actually to lose consciousness at the time of the movement and the attack was described as being "head nodding" in type. The true origin of the attacks was suggested, however, by the fact that the initial thrusting extension which the child showed was constant and sufficiently severe on a number of occasions to bruise the mother's head and neck severely.

It is interesting to note that the three children showing these atypical attacks all showed other epileptic manifestations, grand mal and petit mal in one, grand mal alone in one, and petit mal alone in one.

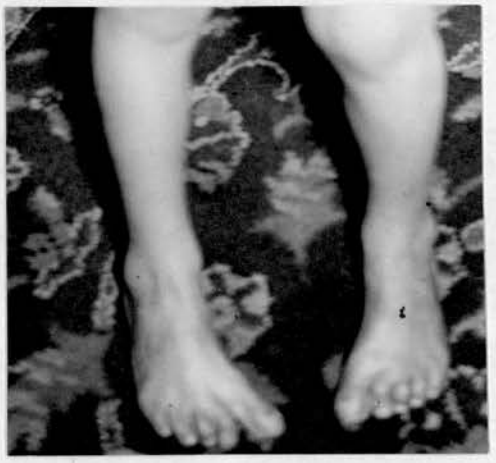
The success of therapy in epilepsy associated with diplegia appears to be rather poor. Though the frequency of attacks, whether grand mal or petit mal, appears to be reduced, only four cases ceased to have attacks while on therapy, and there is nothing to indicate that this was a direct result of therapy.

Other skeletal abnormalities in diplegia.

Dislocation of the hip was encountered in four patients with severe paresis. Three of these were tetraplegic and one triplegic. In one case of tetraplegia the dislocation was bilateral. The dislocation appeared in one between two examinations, between which the degree of rigidity of the lower limbs had very markedly increased. The acetabulum was noted to be poorly for med in two cases, but it seems quite clear that the dislocation is secondary to the extremely powerful pull of the hamstrings in diplegia.

Two cases of diplegia were encountered in which arthrogryposis was present, moderately severely in one and slightly in another. It is interesting to correlate the common positions assumed by the limbs in arthrogryposis to those in severe tetraplegic paresis.

A degree of talipes equino varus, evident at birth and not /



Case I40.Diplegia with ~~bilateral~~ congenital bilateral talipes equino-varus.

not associated with arthrogryposis was noted in 3 cases of diplegia. One case was paraplegic and two were triplegic. In all the cases the talipes was bilateral, though asymmetrical in severity.

As in hemiplegia it seems probably that there is some aetiological connection between congenital talipes equino-varus deformity and diplegia. (Freud, 1897).

Other slight abnormalities of the limbs were found. Syndactyly of the second and third toes occurred in a number of diplegic patients. The hands and feet appeared to be abnormally small in a high proportion, the fingers tending to be abnormally equal in length, soft, hyperextensible and poorly formed.

Other abnormalities.

Congenital heart lesions were encountered in two patients. One was thought to have a patent intra auricular septum and one a patent intra ventricular septum.

Naevie were encountered in nine patients, a finding of interest in view of the recent work on the relationship between haemangiomata congenital eye defects and cerebral abnormalities of

One patient with familial history of neurofibromatosis showed a rather muddy yellow skin with patches of brown pigmentation.

Chapter 3.

Diplegia with cerebellar ataxia.

12 cases were classified as suffering from diplegia with cerebellar ataxia. 7 were males and 5 were females. They were placed in a different category from cases of diplegia without ataxia for a number of reasons. Some of these have been discussed previously in the section dealing with classification. The history of those with and those without ataxia was different. Few ataxic cases are premature. Only one of the patients in this series with diplegia and cerebellar ataxia had a birth weight of less than 5 lbs 8 oz. The importance of intracranial birth injury seemed to be much greater in those with ataxia. No cases showing ataxia had dystonic attacks. A high proportion had a history of familial mental and neurological disorder.

The clinical findings were different. Associated movements were invariably flexor in type, not extensor. The tendon jerks were always much increased and the increase of tone in the muscles was much less marked than in the majority of cases of diplegia without ataxia. Rigidity, except when it was the result of contracture, was not encountered in patients showing ataxia. What increase of tone was present was invariably spastic in type. In most of the cases poor co-ordination was more in evidence than was actual loss of power in the limbs. Some of the patients are similar to those described as 'diataxia cerebralis' by Ramsay Hunt. 1918.

It might be argued that the category of diplegia with

ataxia might have been better classified as a subgroup of diplegic paresis with diplegia without ataxia as another subgroup. But it was felt preferable to emphasise the rather marked differences between these types of diplegic paresis rather than to minimise them.

Further classification of cases of diplegia with ataxia.

All the cases with diplegia and ataxia showed affection of one or both upper limbs, 9 being triplegic and 3 being tetraplegic. Cases were further classified according to the severity of their functional motor impairment which was the result of the ataxia and the paresis in association. The effect of the ataxia was to render the degree of functional impairment due to paresis of the limbs much more severe than it would otherwise have been. In slight cases the effect was merely to make the gait and the child's handling somewhat clumsy. In moderately severe cases the performance of some normal activities was made very difficult, and in severe cases activities were limited, and those that were possible were grossly clumsy. The classification of patients with diplegia and ataxia into mildly affected, moderately severely affected and severely affected is shown in table XXIV.

Five of the patients with diplegia and cerebellar ataxia were females and seven were males.

Clinical findings in diplegia with ataxia.

As in the other groups of patients discussed the clinical findings will be considered firstly from the point of view of the abnormalities found in the limbs themselves, and secondly

Table XXIV.

The severity of the impairme t of voluntary movement and the extent of the paresis in I2 patients suffering from diplegia and ataxia.

Extent of paresis. \ Severity.	Mild.	Moderately severe.	Severe.	Total.
Triplegic.	3	4	2	9
Tetraplegic.	I	0	2	3
Totals.	4	4	4	I2.

the associated abnormalities in the patient impairing his ability to use them.

The paresis.

The paresis was triplegic in distribution in nine cases and tetraplegic in three. The severity of the paresis was not parelleled by the severity of the ataxia. The paresis was more marked in the legs than the arms in all cases. In all, voluntary movements of the limbs were more impaired in the peripheral than the proximal parts of the limbs. In particular fine finger and toe movements were limited and weak. Because of the combination of cerebellar involvement with spastic paresis rapid movements of the fingers and movements requiring much muscular co-ordination were poorer than would be expected in diplegic cases not complicated by ataxia.

As in hemiplegic paresis the movements which tended to suffer most in the upper limbs were thumb and finger extension and abduction, wrist extension and supination of the forearm. In the legs movements of the toes, dorsiflesion of the foot were the most severely affected. In severe cases weakness or limitation of extension of the elbows and knees was evident and abduction of the shoulders and hips was also affected.

As has been noted rigidity due to contracture, was a feature of diplegia with ataxia though rigidity due to alteration of muscle tone control itself was not. The increase of tone evident in cases showing ataxia was very variable in degree, in different cases. In those in which the paresis was slight and ataxia relatively severe hypotonia was evident. In those

with marked paresis spastic increase of tone was evident. In two cases ankle clonus was associated with the typical pendular knee jerks of cerebellar disease. Because of the hypotonic element in the cases of diplegia with ataxia the tendon jerks showed very marked exaggeration.

Contracture of the limbs was much less marked than in cases of diplegia which did not show ataxia. When it occured it was invariably in flexor positions, and of "hemiplegic" type.

In all the patients with diplegia and ataxia the plantar responses were extensor bilaterally.

The ataxia.

The ataxia varied in severity from mild to severe in different cases. In two cases it was predominantly right sided, in six predominantly left sided and in four it was apparently equal in severity bilaterally.

The ataxia was manifested in inco-ordination was shown as clumsiness, only on voluntary movement. In severe cases the inco-ordination was so severe that handling of objects was grossly impaired, writing was impossible and small movements very difficult. Of the eight moderately severe and severe cases of diplegia and ataxia none was able to do his own buttons or laces, and in some of these the paresis was relatively slight. The ataxia was most evident clinically in the broad based stamping gait of the child and in the tendency to passpoint on testing. In those with asymmetrical ataxia the tendency to deviate to one side on blindfold or backwards walking was marked.

The Romberg test was found to be consistently positive only in cases with severe ataxia. In others the compensation for the cerebellar deficiency was good enough for it to be normal. The finger nose and finger to finger tests together with backwards walking with the eyes shut were found to be far more reliable ways of detecting ataxia.

The gait was affected somewhat differently in some cases than in others. In some the gait was on a very broad base and was stamping in type.

Male born 1945. Case 27.

A case of moderately severe triplegia with predominantly left sided cerebellar ataxia.

The second child of healthy parents. Mother aged 31 at the time of birth. Previous child healthy. One previous miscarriage.

She was well in her pregnancy with the patient. Labour occured at term, lasted $8\frac{1}{2}$ hours and delivery was spontaneous in hospital. The mother had a post partum haemorrhage.

The child birth weight was 9 lbs 2 oz and he seemed to be normal. The neonatal period was normal except that there was a weight loss of 8 ounces, which could not be explained and he was kept in hospital for two weeks, at which time he had regained his birth weight.

He was rather a quiet infant but no abnormality was noted except that his milestones were slow. He sat with support at the age of 10 months. He stood at 18 months but was able to walk without support only at the age of 2. For six months

Case 27.Patient with diplegia and cerebellar ataxia.The latent weakness of the left arm is shown by the slight flexion of the fingers and elbow and the pronation of the forearm.He was unable to stand steadily with the feet any closer together than is shown in the photograph.

before this he had seemed very uncertain and unable to "let himself go". The mother wondered why his muscles were so flabby and he was "so collapsible".

He used his right arm at the age of about 6 months. The left was not used until the age of 10 months. His handling was bilaterally very clumsy. He said his first words at the age of 2.

When he first began to walk he fell a great deal and seemed to be very unsteady. He usually fell flat on his face or on the left side. His knees were never without sticking plaster. The falling became less marked about the age of 3.

He went to nursery school at the age of 3 and kinder-garden at the age of 5. At school his reading and writing have been very slow in developing and he has showed a great tendency to reverse small words and letters and to put them horizontally instead of vertically and vice versa. Intelligence on Terman Merrill 105. Weschler Performance test gave an I.Q. of 67, largely due to his poor handling and grasp of spatial concepts.

On examination at the age of 8, his height was 49½". Head circumference was 21½". There was no asymmetry of the head, limbs or trunk.

He was alert and intelligent. His speech appeared to be normal, and of good content. There was slight emotional overaction of the left face and leg on voluntary movements though the face was symmetrical at rest.

The power of the upper limbs was fair. It was less good

on the left than on the right and supination of the left forearm, extension of the fingers and elbow was weaker than on the right. The lower limbs showed greater weakness bilaterally. Dorsiflexion of the feet was very weak and movements of the toes were very poor. Co-ordination was more impaired than power in all the limbs. Rapid and fine finger and toe movements were very poor on the left and impaired on the right. The right finger nose and heel knee tests were impaired but not badly performed. The left finger nose and heel tests were very poorly performed. There was a constant tendency to overreach to the left when he attempted to grasp objects, especially with the left hand.

He could not do buttons or laces because of the clumsiness of the left hand. Movements of the left hand were accompanied by slight athetosis of the fingers, especially the ring and middle fingers, which was not evident on the right.

The tone of the legs and left arm was normal. The tone of the right arm was hypotonic. The biceps, triceps and supinator jerks on the left were exaggerated to a surprising extent. The knee and ankle jerks were bilaterally very marked. The plantar responses were bilaterally extensor.

His gait was abnormal. The legs were held widely apart and the feet were almost stamped on the ground as he proceeded. There was no equinus position of the feet. There was some associated flexion of the left arm when he walked or used the right arm voluntarily. There was considerably swaying of the body from side to side as he walked. When blindfold or made

to walk backwards he deviated constantly to the left. When descending stairs he tended to keep to the left side and was very uncertain, slow and careful. No loss of cutaneous sensibility was demonstrated.

In others the spasticity was more marked and the gait was more typically diplegic, than spastic in type.

Female born in 1943. Case IOI.

A case of moderately severe triplegia with cerebellar ataxia. The mother suffers from moderately severe tetraplegia with ataxia and is mentally defective. States that there is a sister like her.

Patient was born when the mother was aged 38. She was well in pregnancy. States delivery was at term in hospital. The birth weight was $3\frac{1}{2}$ pounds and there was delay in her being discharged. Milestones uncertain but she walked holding onto furniture from the age of about 2 until the age of 3 when she began to walk without support. When she did so she was very unsteady and fell a great deal. She walked on her toes,"like the rest o' us". She fell forward, not to either side more often than the other. She fell a great deal until the age of 7 when she began to attend school for the phycially handicapped. There she had made rather poor progress. I.Q. 70.

Examination when aged 10. Her height was $53\frac{1}{2}$". Head circumference was $20\frac{1}{2}$". There was no asymmetry of the limbs or the trunk or skull. She was slow and dull, but able to

respond to simple questions and commands.

Her speech showed grossly defective pronunnciation of consonants. There was a slight left facial paresis of upper motor neurone type, evident only on movement.

There was defective power in the left upper limb compared to the right and extension of the fingers and supination of the forearm, especially were weak. The right upper limb appeared to show normal power. The lower limb showed much reduction in power, dorsiflexion of the feet was limited bilaterally. The toes could not be moved voluntarily. Full extension of the knees and hips was weak.

There was poor co-ordination of all limbs. The finger nose tests were very inaccurate and the heel knee tests grossly unsteady. There was bilateral falling away of the hands on her being asked to maintain them extended and held forwards.

There was increase of tone of moderate degree and spastic type in the left upper limb and moderately severe bilateral spastic increase of tone in the lower limbs. The biceps, triceps and supinator jerks in the left upper limb were moderately increased compared to the right. The knee and ankle jerks were markedly increased bilaterally and ankle clonus was present. The plantars were extensor. When she walked or used the right arm there was a tendancy to flexor associated movements in the left arm.

The gait was spastic in type with the feet bilaterally in the equinus position and a marked tendency to scissoring.

The hips and knees were maintained in a somewhat flexed position and she leant forwards as she walked. She rolled about from side to side as she walked, and showed very grossly defective sense of direction when blindfold. There was a tendency to festination.

The Romberg test resulted in her falling flat on her face. No defects of superficial sensation could be demonstrated.

In most cases a history of many falls in the first two or three years after learning to walk was elicited. Frequently the child had found going down stairs impossible for several years after he had learnt to go up, and three cases still crawled down stairs back to front when they were over 5 years old. Later on the side of the stairs to which the child tended to hold when he came down stairs often indicated the side of the ataxia. All the cases with ataxia of any severity showed much less confidence in the dark than normal children. This was presumably due to the fact that their means of visual compensation for their attaxic disorder was rendered impotent.

Other findings in the limbs.

Apparent shortening in the limbs was not a feature of diplegia with cerebella ataxia. Unfortunately the number of cases is too small to confirm the impression that generalised dwarfing does not occur, but four cases were over the age of 12 years and were within normal limits for height.

The feet tended to be colder than the hands in cases of diplegia with ataxia, as in diplegia. The marked vasomotor changes found in hemiplegia were not found however.

No disturbances of superficial sensation or in joint or vibration sense were discovered in any of the patients with diplegia and cerebellar ataxia.

Intellectual impairment.

Five of the cases were within the normal range of intelligence and two were considered ineducable. That the combination of spastic diplegia and ataxia was a particularly severe handicap from the educational point of view is indicated by the finding that only two of the five children with normal intelligence were in normal schools. This is shown in tables XXV and XXVI .

Unfortunately the numbers of cases in the survey are too small for a comparison to be made between the intelligence and schooling of cases showing ataxia alone, diplegia alone and the combination of diplegia and cerebellar ataxia.

Speech disorder.

The numbers of cases showing various types of speech disturbance are shown in table XXVII. It will be seen that 10 of the patients suffered from speech abnormality sufficient to lead to some difficulty in comprehending what they said. Seven patients showed dyslalia. Two of these patients also showed scanning speech. The severity of the dyslalia varied from severe in which the speech was found difficult to understnad even by relatives, to slight where relatively little difficulty in comprehension was encountered. One patient showed marked scanning of speech as an isolated speech defect. No cases of stutter or marked hesitation in speech were

Table XXV.

The intelligence quotients of I2 cases of diplegia with ataxia.

Intelligence quotients. Severity of paresis.	II5 or more	I00-II4.	85-99.	70-84.	55-69.	Under 55.	Untested.
Mild.	0	0	0	I	2	I	0
Moderately severe.	0	2	I	0	0	I	0
Severe.	0	0	2	0	0	0	2
Extent of paresis.							
Triplegic.	0	2	3	I	I	2	3
Tetraplegia.	0	0	I	0	I	0	I
Totals.	0	2	3	I	I	2	3

Table XXVI.

Schooling of I2 patients with diplegia and ataxia.

School.	Triplegic cases.	Tetraplegic cases.	All. cases.
Normal school.	2	0	2
Schools for physically handicapped.	3	I	4
Schools for the mentally handicapped.	I	I	2
Ineducable.	2	0	2
Not yet at schools.	I	I	2
Totals.	9	3	I2.

encountered.

Visual defect.

One patient showed very pale, clearly demarcated optic discs on retinal examination. The appearance suggested optic atrophy. In spite of the absence of apparent refractive error his visual acuity was only J4 bilaterally.

Two patients had very severe myopia.

No cases showing amaurosis or field defect to confrontation were encountered.

Other cranial nerves.

Strabismus. Three patients showed strabismus. Two showed bilateral abducent paresis, in one complicated by unilateral third nerve weakness and impaired ability to close the eyes voluntarily. One case showed a unilateral abducent paresis. No cases showing clinically recognisable supranuclear paresis were found.

Facial Paresis. Five patients showed facial paresis of upper motor neurone type. Two of these were tetraplegic and three were triplegic. Two were considered to be cases with mild affection of the limbs, one with moderately severe affection and two were severely affected. One case, suffering from severe triplegic paresis with cerebellar ataxia showed a very interesting bilateral upper motor neurone facial paresis. The same variations in the manifestations of the paresis found in hemiplegic and diplegia patients at rest and on emotional and voluntary movements were also encountered in cases of diplegia with ataxia.

Table XXVII

Speech defects in I2 patients with diplegia and ataxia.

Extent of paresis.	Number of cases.	Dyslalia. Mild.	Moderately severe.	Severe.	No comprehensible speech.	Scanning.	Total with def
Triplegia.	9	3 x	I	2 x	2	2 xx	8
Tetraplegia.	3	0	I	0	0	I	2
Totals.	I2	3	2	2	2	3	IO.

x marks patients with dyslalia and scanning speech.

Swallowing and tongue involvement.

One patient, ~~case~~ referred to above, showed evidence of difficulty in swallowing. Swallowing was infrequent and drooling excessive. The same boy also showed limitation of voluntary movements of the tongue.

Epilepsy.

Three patients showed epileptic phenomena. Two of the patients were triplegic and two tetraplegic TableXXVIII One case was slightly, one moderately severely and one severely affected by diplegia and cerebellar ataxia. None showed grand mal fits. One showed Jacksonian attacks and petit mal. One had petit mal and saalam attacks with loss of conciousness for about one minute in the attacks which occured once or twice every day. The third case showed petit mal anly.

The petit mal attacks started at the age of 5 months in one case and between the ages of 1 year and three years in the other two.

Behaviour.

One case showed hyperactive behaviour similar to that noted in hemiplegic patients and described in the section discussing the findings in hemiplegia.

Other findings.

One case showed congenital hydrocephalus, probably the result of birth injury followed by haemorrhage which had been treated by third ventriculostomy and appeared to be controlled. The head circumference when the child was examined at the age of 4 years was 24.6. inches.

Table XXVIII

Epilepsy in 12 patients with diplegia and ataxia.

Type of epilepsy.	Grand mal.	Petit mal.	Other.	Totals. Number of cases.
Extent of paresis.				
Triplegia.	0	2 x	I x	2
Tetraplegia.	0	I x	I x	I
Totals.	0	3	2	3

x One patient markedso,had petit mal and other fits.

One case showed scattered areas of brown skinpigmentation over the back and chest. The patches were illdefined and irregular in shape. Their size varied from a few millimetres in diameter to 4 by 5 inches.

Chapter. 4.

The Clinical findings in 15 cases of ataxia.

Fifteen children with cerebral palsy were classified as suffering from ataxia of the limbs as their predominant disability. In 14 of the patients the ataxia was cerebellar in type. In one the ataxia was mixed cerebellar and vestibular in type, the result of tuberculous meningitis treated with streptomycin.

Of the fifteen cases nine were males and six were females. The severity of their ataxia, its distribution and the clinical findings in the patients varied greatly. All the cases showed impairment of co-ordination of the limbs however, and in none was paresis of the limbs any more than a slight disability. Power of the limbs tended to be within normal limits. Use of the power was defective.

The extent of the ataxia and its severity.

The ataxia was central in type in 9 cases as shown in Table XXIX. In these the child was more or less unsteady on the upright position but tended to deviate to one side no more than to the other. The severity of the ataxia of the limbs was not necessarily quite so symmetrical and most of these cases showed somewhat greater inco-ordination of voluntary movement one one side than on the other, though all the limbs were affected to some degree.

In six patients the ataxia was predominantly unilateral, the child tending to deviate and passpoint to one side more than the other. Two cases showed predominantly right sided

Table XXIX

The extent and severity of ataxia in 15 patients.

Severity of ataxia.	Slight.	Moderately severe.	Severe.	Totals.
Direction of ataxia.				
Predominantly right.	I	I	0	2
Predominantly left.	I	3	0	4
Bilateral.	2	4	3	9
Totals.	4	8	3	15.

ataxia and four left sided ataxia. Though the limbs on the affected side in these patients showed inco-ordination on voluntary movement, the opposite limbs also showed some impairment of co-ordination, less marked in degree.

Four patients were classified as suffering from ataxia of mild degree. In these normal activities were possible though they might be somewhat clumsy and motor milestones retarded to some extent. Eight patients were classified as having ataxia of moderately severe degree. In these patients some normal activities were impossible and others impaired, but the ataxia was not severe enough to prevent independent gait and writing. Severe cases showed great impairment of their ability to perform normal tasks. Their gait was grossly unsteady, or they had no independent walking and their manipulations were extremely clumsy so that their writing was extremely defective. They required help in dressing.

The manifestations of ataxia.

In the majority of cases the most evident manifestation of underlying ataxia was abnormality of the gait rather than gross unsteadiness in the erect position. The legs tended to be wide apart, the feet to be stamped to the ground and the steps to be small. In cases in which the ataxia was more marked to one side than to the other the shoulder of the more affected side tended to be lower and more forward than that on the affected. In two cases the difference in the levels of the shoulders, was as much as 2". The back showed a scoliosis in the thoracic region towards the less affected side and the

head was tilted towards it, with a scoliosis in the cervical region towards the affected side.

That the appearance of steadiness which most of these children showed was the result of walking on a broad base and compensating by visual means for their ataxia, this could easily be demonstrated. When requested to walk along a straight line and to place their toes upon it a marked tendency to fall was often evident. When asked to walk with the eyes closed or blindfold the majority of the cases were very unsure and tended to fall to one or other side. This was especially evident when the child was asked to walk backwards blindfolded, and ataxia was much more evident on this test than on the Romberg test, which was found to be rather unreliable. It was positive in only 5 cases, where as walking backwards blindfold revealed ataxia in all those in the series.

The majority of cases with ataxia showed delay in walking; the average age of achieving independent walking in 10 cases thought to be of congenital origin was 2 years 3 months. Three cases walked independently only after the age of $3\frac{1}{2}$ years. There tended to be a period of three or 4 months during which the child would walk holding on to furniture but not without support. When independent walking was achieved most children fell extremely frequently, and the knees of most cases of ataxia bear numerous scars the result of this early marked unsteadiness. As a somewhat later stage the patients have the greatest difficulty learning to manage stairs and especially to descend them. The side of the stairs to which the child holds in his

descent was found to be a good guide to the side on which the ataxia was more severe. Two cases with bilateral ataxia still descended stairs hind end foremost, on all fours at the age of 6.

As when they were blindfolded, cases of ataxia tended to be uncertain when it was dark. Though predominantly unilateral cases could walk remarkably straight with their eyes open and their legs wide apart. As soon as they were deprived of the use of their eyes very marked deviations to the affected side resulted in all except those who were slightly affected. In all patients with ataxia however there appeared to be a very poor sense of direction and in a school for spastics ataxia cases may frequently be seen to go to the wrong end of corridors after three or four years of residence. Their poor sense of direction was also shown in the loss of direction shown in the writing of cases of ataxia, and its tendency to stray from the horizontal on the page.

On testing all of those with predominal unilateral ataxia showed pass pointing towards the ataxic side. When the arms were held forwards and extended the limb on the affected side showed falling away before the opposite arm. In bilateral cases the posture of the limbs was poorly maintained and the falling away was usually less consistently to one side or the other. Similarly cases with predominantly unilateral ataxia tested by the finger to nose to observers finger test showed pass pointing consistently in one direction.

Female born 1941. Case 28.

A case of moderately severe cerebellar ataxia predominantly to the left. The second of three children born to father suffering from chronic nephritis and healty mother. The first child is healthy. The third child has a spinabifida.
The patient was born when the mother was age 26 in hospital. She had preeclampsia in the later months of pregnancy. This was not severe.
Premature rupture of the membranes occured 4 or 5 weeks before term and the child was delivered spontaneously after $6\frac{1}{4}$ hours.

The child weighed 5 lbs $2\frac{3}{4}$" at the time of birth and was not asphyxiated. She was noted to be rather feeble, however. She was nursed in a premature nursery for two days and given two hourly feeds.

She was a good quiet infant. She sat with support at the age of 8 months and stood at the age of 17 months with support. She said her first words shortly after the age of 1 year. She used her right hand from the age of about 4 months, has always used the left to a less extent than the right. A squint was noted at the age of 6 months.

She began to walk at the age of about 20 months without support, having held onto furniture for three months before this. She fell a great deal usually backwards or to the left. Her falling persisted until the age of about 7 when it became less frequent. It was only at this age that she meant to manage to go up stairs, holding onto the left banister. She could only come down upright at the age of 10.

She went to school at the age of 5 and has made very poor progress. She reads a little but is unable to write more than her name. "Puts her letters old anyhow" says her teacher. I.Q. 65.

On examination at the age of 12. Height $53\frac{3}{4}$". Head circumference $19\frac{1}{2}$". There was no asymmertry of the head or limbs. A right scoliosis of the thoricic and lumbar spine was present.

She was rather slow in responses to commands and questions. Her speech was poor in content showed gross impairment of pronounciation. Consonant were very poorly pronounced. Some scanning was apparent. When she was excited her speech was incomprehensible.

There was an eccentricity of the right pupil. She was hypermetropic. The eye movements were bilaterally limited by abducent paresis.

The power of the limbs appeared to be normal. No paresis was evident. Co-ordination of the limbs was very poor however. Fine and rapid movements of the left fingers were impossible and they were rather poor on the right. The toes could be moved only very sluggishly. The finger nose test on the left was grossly inaccurate. That on the right was somewhat impaired. The heel knee tests were impaired more on the left than the right. There was marked passpointing to the left on testing. This was evident in all limbs but more marked in the left limbs than in those on the right.

The tone of the limbs was less than normal. There was

greater hypotonia on the left than the right. The tendon jerks were normal and the plantar responses were flexor. No associated movements of the limbs were evident.

The gait was abnormal. The walking base was broad. When she was blindfold she deviated markedly to the left whether walking forwards or backwards.

No abnormalities were demonstrated in superficial sensation.

Cases with bilateral ataxia showed grossly defective sense of position rather than true consistent passpointing in any one direction. The result of ataxia in the upper limbs was that the children were unable to reach for objects without looking where they were putting the hand. This may not sound a particularly severe disability in itself. But it is only necessary to watch an ataxic child drawing and rubbing out what he had drawn in the wrong direction, then look and reach for his pencil, then look and reach for his rubber to realise the time he wastes in his fruitless and abnormal activity.

Disturbances in the limbs in the patients with ataxia.

Clumsiness of movement was invariable in patients with ataxia. It affected fine and rapid movements, and therefore movements of the distal parts of the limbs, to a greater extent than coarse movements. In particular fine and rapid movements of the fingers were impaired and of the 10 congenital cases buttons and laces could only be managed without help in six, and in none of these before the age of 5. The clumsiness of movement tended to be noted in congenital cases at an early age

and in 7 the child was stated to have been clumsy with one or both hands before the age of 1 year. The clumsiness was marked only on the more affected side in the cases showing predominantly unilateral ataxia, but on examination it was usual to find that some degree of dysdiadochokinesia was also present in the fingers of the opposite side.

Intention tremor of moderately coarse type was present on voluntary movements of the hands and feet on the affected side in two patients with predominantly unilateral ataxia. They were absent on the opposite side. In both cases well marked hypotonia was present on the side to which ataxia was evident.

Well marked hypotonia was present in five patients, three of them cases of predominantly unilateral ataxia and two cases of bilateral ataxia. All the cases of unilateral ataxia showed unilateral hypotonia, on the side to which the ataxia was evident. One of the cases of bilateral ataxia showed hypotonia only on one side while the other showed generalised hyptonia.

The reason why a higher proportion of cases did not show hypotonia is that many had other minor neurological abnormalities of upper motor neurone type, resulting in slight spastic increase of tone replacing hypotonia. In two patients no neurological abnormality could be discovered other than ataxia however and tone in the limbs was normal. One of these cases were severely and one slightly affected by ataxia. The types of upper motor neurone lesion encountered which gave rise to slight increase in the tone of the limbs were hemiplegic in

type in six cases and paraplegic in two cases. These findings are shown in table XXX .

The patients showing hemiplegic increase in tone were somewhat similar in type. Apart from slightly greater impairment of fine and rapid movement of the fingers on the affected side and slight weakness of supination of the fore arm, finger extension and movements of the thumb, paresis was not a striking feature. On voluntary activity of the contralateral arm however, associated movements of the flexor types were present in the hemiplegic upper limb, the hand clenching, the forearm pronating and the elbow flexing slightly. On examination of the tone of the limbs on the hemiplegic side some increase of tone of spastic type was evident. The biceps, triceps, supinator, knee and ankle jerks were increased out of proportion to the degree of increase of tone. The knee jerks in some cases were exaggerated and seemed to show a mixture of spastic increase of power and pendular increase in range of the response to stimulation.

Three of the patients showed extensor plantars on the hemiplegic side. In three cases the responses were equivocal or flexor. Four patients showed cold hands and feet on the side affected by hemiplegic increase in tone.

One case showing bilateral ataxia with mild hemiplegia showed slight athetosis of the fingers on the affected side. Only one patient showed apparent shortening in the hemiplegic upper limb of more than $\frac{1}{2}$" and in her case it was 2/3". Paresis of her affected arm was very slight, and the limb was

Table XXX

Associated neurological abnormalities in the limbs in I5 patients with ataxia.

Severity of the ataxia.	Number of patients.	Increase of muscle tone.		Involuntary movement.		Hypotonia.	
		Paraplegic.	Hemiplegia.	Athetosis.	Tremor.	Unilateral.	Bilate
Mild.	4	0	I	0	0	2	0
Moderately severe.	8	I	4	I	2	I	I
Severe.	3	I	I	0	0	I	0
Totals.	I5	2•	6x	I	2	4	I

• One with bilateral Babinski signs.

x Three with unilateral Babinski signs.

useful.

Boy born in 1944. Case149.

A case of severe bilateral cerebellar ataxia.

Two of the mother's brothers had meningitis in childhood and are now well. The mother had meningitis in childhood. The patients elder brother died of meningitis age 3. Two younger children are well.

He was the second born child or four. The pregnancy was normal and labour was at term. Delivery was spontaneous after 1½ hours. He seemed normal at birth. The weight was 7 lbs 6 oz.

At the age of 5 weeks he became fretful, cried a lot and seemed fevered. Following a generalised convulsion which lasted for some hours he was transferred to hospital suffering from meningococcal meningitis, a diagnosis reached by the mother before the doctor.

He was discharged at the age of 4 months with a squint and an asymmetrical face though otherwise he seemed well.

He sat at the age of 7 months and stood holding on at the age of 9 months. He said his first words at the age of one year. At the age of 15 months he began to walk a few steps occasionally but seemed very uncertain and it was 18 months, 9 months after he had stood holding on, that he was able to walk without support. When he did so he was very unsteady and fell a great deal, either forwards or backwards. Falling was frequent until the age of 5 when it became less marked.

He used his hands about the normal age but handling has always been clumsy and he is very liable to drop things. He was unable to do buttons or laces at the age of 9.

At school he has made good progress but reading is in advance of writing.

Examination aged 9. Height 47". Head circumference 19½" The child was intelligent and responded briskly to commands and answered questions accurately and well. The speech was scanned.

The vision of the left eye was defective.

There was a left facial palsy of lower motor neuron type.

There was a complete deafness of the left ear.

The power of the limbs appeared to be normal and no paresis was demonstrated. Co-ordination of voluntary movements was impaired bilaterally, in upper and lower limbs. Fine and rapid movements of the limbs were very poor. He was unable to pick up small objects and could not do laces or buttons. When the upper limbs were outstretched forwards there was a very rapid falling away bilaterally.

The finger nose tests were grossly inaccurate but pass-pointing to one side consistently could not be demonstrated.

The tone of the left limbs was normal. There was a very slight increase of tone in the limbs on the right of spastic type. The biceps, triceps, supinator, knee and ankle jerks definitely pendular. The plantar responses were flexor.

The gait was ataxic. The walking base was broad and the feet stamped down. There was no equinus and associated movements of the limbs were not marked.

When blindfold he walked all over the room attempting to walk in a straight line. The Romberg test was strictly negative though he was very easily pushed to one or other side.

No abnormalities of superficial sensation could be demonstrated.

Two cases showed slight paraplegic increase of tone of spastic type in each case. Both were cases of moderately severe bilateral ataxia, and both were cases of congenital hydrocephalus. In neither case was the increase of tone in the lower limbs more than slight, and since paresis of the limbs could not be demonstrated it was felt that they should be justified as cases of ataxia and not deplegia with cerebellar ataxia. One case showed extensor plantar responses and one extensor plantar responses bilaterally. One of the patients showed a lower temperature in the feet than the hand. Neither case showed apparent dwarfing.

Apart from the case previously referred to no asymmetry of the limbs was evident. No cases other than those already mentioned showed differences of temperature in the limbs. Neglect of the affected limbs was not a feature of ataxia.

One case of ataxia following tuberculous meningitis showed partial superficial hemianaestheia to all modalities and impairment of joint and position sense. The patient showed predominantly unilateral ataxia of moderate severity associated with slight hemiplegic increase of tone, on the same side as that on which the sensory impairment was present.

Table XXXI.

The intelligence quotients of 15 patients with ataxia.

Intelligence quotient.	115 or more.	100-114	85-99	70-84	55-69.	Under 55	Untested.
Number of patients.	0	1	3	6	3	0	2

Table XXXII.

The schooling of 15 patients with ataxia.

School.	Number of patients.
Normal schools.	2
Schools for the physically handicapped.	7
Schools for the mentally handicapped.	3
Ineducable.	0
Not yet at school.	3
Total.	15

Intellectual impairment in ataxia.

The intellectual impairment in the cases of ataxia as measured by different tests given in different circumstances by different testers are shown on tables XXXI & II and it will be observed that only four of the fifteen cases had intelligence quotients over 85.

The schooling of the cases of ataxia is shown in table It will be seen that more than half of those over the age of 5 attended schools for the physically handicapped, a reflexion of the severe educational disability that ataxia imposes. This disability is partly the result of the slow manipulation which the patients show, partly the result of the difficulty they have in learning to recognise and form letter, and partly the result of the intellectual impairment.

Behaviour.

Four patients showed overactive behaviour of the type described in the section on hemiplegic paresis. In three the overactivity was marked and in one was less evident. Three of the patients showing overactive behaviour showed epilepsy. The overactivity was sufficiently severe in all the cases to make education by normal methods a very difficult and strenuous undertaking for the teacher.

Speech defects in ataxia patients.

It will be seen from table XXXIII that 11 of the 15 ataxic patients showed speech defects. In 8 cases dyslalia of varying degrees of severity was present, in four of the cases accompanied by scanning speech. Scanning speech without apparent dyslalia was evident in thee cases.

Table XXXIII.

Speech defects in I5 patients with ataxia.

Disorder of speech.	Number of patients.
Dyslalia	4
Scanning speech.	3
Dyslalia and scanning speech.	4
All defects.	II.

Table XXXIV.

Cranial nerve involvement in I5 patients with ataxia.

	Number of patients.
Visual defects.	2
Strabismus (unilateral.	I
Strabismus (bilateral.	4
Nystagmus.	2
Facial paresis (upper motor neurone.	4
Facial paresis (lower motor neurone.	2
Deafness (unilateral.	I
Deafness (bilateral.	I
Tongue involvement.	I

In all the cases the defect was sufficiently severe to make comprehension of what was said difficult to some degree. In one of the cases of dyslalia and in two with scanning speech and dyslalia the speech was grossly defective.

Visual defects.

One patient with severe predominantly unilateral ataxia showed unilateral optic atrophy with complete unilateral amaurosis. The opposite eye was unaffected. One patient with bilateral ataxia of moderately severe degree and mild left hemiplegia showed complete left homonymous hemianopia, and some impairment of vision in the remaining half field in the lower quadrant bilaterally. One patients showed severe bilateral astigmatism.

Cranial nerve involvement in cases of ataxia.

These are shown in table. XXXIV.

Strabismus.

Unilateral abducent paresis was present in one case and bilateral abducent paresis in four patients. In the latter the abducent paresis was asymmetrical in severity in three of the patients and symmetrical in one. The patient with unilateral abducent paresis also had amaurosis of one eye. No cases of third nerve involvement were encountered.

Nystagmus.

Two cases had nystagmus. One of these showed homonymous field defects, and nystagmus of a coarse type in all directions. She had bilateral moderately severe ataxia. The other case also showed moderately severe bilateral ataxia.

Nystagmus was present only on gaze to the right and was slow and swinging in type.

Facial involvement.

Five of the 6 patients with mild hemiplegia associated with their ataxia showed facial paresis of upper motor neurone type. In none of the cases was there asymmetry at rest but slight lag was evident on voluntary movement. In four of the cases this lag was also evident on emotional movement, but one showed overaction of the face on emotional movement, and lag on voluntary movement.

One patients showed facial paresis of lower motor neurone type which followed damage to the facial nerve suffered during meningitis.

Hearing.

One patient was totally deaf in one ear as a result of meningitis. One patient was totally deaf as a result of tuberculous meningitis treated by streptomycin. He also showed absence of vestibular response bilaterally on the caloric tests.

Swallowing.

None of the ataxic patients showed apparent difficulty in swallowing at the time of examination.

Tongue.

One patient with moderately severe predominantly unilateral ataxia, slight hemiplegia and partial hemianaesthesia showed a tendency for the tongue to deviate to the right when central protrusion was attempted.

Epilepsy.

Three patients with ataxia showed grand mal elipsy. In two the ataxia was predominantly unilateral, and in one bilateral. One of the cases was severely affected, one moderately severely and one slightly. One of the cases of grand mal also suffered from petit mal attacks.

One of the cases of grand mal had suffered from attacks once or twice a week from the time of birth to the age of 3, at which age they had ceased. The attacks had been accompanied by loss of conciousness, generalised twitching of the limbs and incontinence.

One girl with moderately severe predominantly unilateral ataxia slight hemiplegia and hemianaesthesia following tuberculous meningitis showed grand mal attacks which occured once every week on average. They were accompanied by loss of conciousness, incontinence and generalised twitchings of the limbs. The attacks lasted approximately 5 minutes on most occasions but conciousness had been lost for as long as half an hour in some. The attacks were little affected by therapy. The patient with petit mal, in addition to grand mal showed attacks of the latter type over 3 - 4 months. They were severe in type with generalised twitching of the limbs and loss of conciousness for up to 3 hours. Her petit mal attacks occured many times a day and were observed at one examination to have occured 11 times in one and a half hours in spite of drug therapy.

One patient with moderately severe bilateral ataxia had 7 attacks of epilepsy between the age of 14 months and 4 years

which were of Jacksonian type and which started in his right hand, on the side affected by slight hemiplegia. Following the administration of phenobarbitone no attacks had been present for 18 months at the time of examination.

Hydrocephalus.

Two patients showed hydrocephalus of congenital type. In one the hydrocephalus had become arrested spontaneously. The occipito frontal circumference was 25". One case had had third ventriculostomy and the head circumverence was 21.3" at the age of 3.

Other findings.

One patients, without apparent visual defect showed a curious abnormality of the vessels in one retina. They were abnormally tortuous towards the temporal pole, almost angiomatous, but X-ray of the skull showed no abnormality and other angiomata were not discovered.

One patient had a congenital heart defect, thought to be a patent interventricular septum.

Chapter 5.

The findings in 17 cases with dyskinesia.

The term dyskinesia describes cases showing impairment of voluntary activity as a result of involuntary movements of various types. Perlstein 1952. These types are very greatly in different patients and their classification is very difficult. 17 patients, 7 male and 10 female were classified as suffering from dyskinesia.

Classification of types of dyskinesia.

There are two practical methods of attempting to classify cases of dyskinesia. The categories may be made very numerous in an attempt to cover the numerous variations in clinical picture encountered, or these variations may be ignored and a much more approximate classification attempted. An example of an attempt to cover all the clinical types isfound in one modern American classification which comprises 17 sub groups with different clinical pictures. Unfortunately, in spite of its complexity the classification still fails to describe a large number of the cases very fully.

In this survey a much simpler classification is attempted. It is acknowledged that the complexity of the clinical findings will defeat any attempt at full classification. Therefore the categories are based on the type of involuntary movement exhibited by the child, the distribution of the movembents in the limbs and the severity of functional impairment that results from them.

The types of dyskinesia encountered in the survey.

It will be seen from table XXXV that five types of

Table XXXV.

The types and distribution of involuntary movements in 17 patients with dyskinesia.

Distribution of movements. Type of movement.	Hemiplegic.	Triplegic.	Tetraplegic.	Totals.
Athetosis alone.	0	I	0	I
Choreoid only.	0	0	I	I
Athetoid and choreoid only.	0	4	2	6
Athetosis with tremor.	0	0	I	I
Choreoid movements, athetesis and tremor.	0	0	I	I
Choreoid movements with dystonia.	0	0	I	I
Choreoid, athetoid and dystonic.	0	I	I	2
Choreoid movements with tension.	0	0	I	I
Choreoid, athetoid and tension.	I	I	I	3
All types of involuntary movement.	I	7	9	17

involuntary movements were recognised in the patients. These occured relatively infrequently alone. More often two or more forms of involuntary movement could be recognised in the same patient. Gowers 1874.

The commonest types of movement were choreoid and athetoid in type. They were found in association in 12 patients. Athetoid movements were similar to those encountered in hemiplegic cases except that their severity and extent seemed to be somewhat greater in those suffering from dyskinesia. The wrist and forearm was more often involved in these cases, in addition to the fingers.

Choreoid movements were similar in type to those of rheumatic chorea. In fact a surprising number of patients in the series had had long period of bed rest and salicylates in hospitals, without, needless to say, striking benefit. In contrast to athetoid movements, choreoid movements were more marked in the proximal joints than in the distal. In most patients the choreoid movements were more severe on one side than the other, and when they occured they were liable to affect the face, head and neck, in addition to the limbs. Choreoid movements were almost invariably associated with a marked momentary increase of tone in the limbs affected. Tremor was apparent in two patients in the series. In both cases it was a coarse type of intention tremor.

By tension was understood a sudden increase of muscle tone in the limbs. Usually the increase of tone was most marked in the limbs undergoing voluntary movement but it was also

apparent in the other limbs. It occured in all the cases showing choreoid movements to a greater or lesser degree but in some patients it was such a severe cause of impairment of function that it was felt desirable to make a separate category for them. In these the whole voluntary movement was halted and disorganised by the occurence of tension.

By dystonia was understood involvement of the trunk muscles in the involuntary movement, usually associated with extension of one or both upper limbs. Dystonia was also always associated with sudden increase of the tone of the muscles which resulted in disorganisation of voluntary movement. But in addition the dystonic movement, as in diplegia resulted in further difficulties for the child because of the severe generalised alterations in posture which resulted from quite small range voluntary movements. In the cases of dyskinesia dystonia was never encountered as an isolated finding, but was always associated with choreoid movements.

The development of dyskinesia.

As has been noted, a high proportion of cases of dyskinesia are misdiagnosed and treated as cases of rheumatic chorea, habit spasms, epilepsy, mental deficiency, and, in two cases in the series as cases of progressive encephalitis. The reasons for the frequency of misdiagnosis are that the development patterns shown by the cases of dyskinesis are curious and little known.

None of the cases showed the involuntary movements which were evident later at the time of birth. Motor milestone, holding up the head, using the hands, sitting and standing

were slow in all but one of the patients. The most frequent pattern of development of choreic movements was that when the child was beginning to hold up the head, usually between 6 and 18 months, spasms of the limbs and trunk became evident, most frequently when the child was handled. In three cases a very close association was apparent between the development of the spasms and infections of various types.

Female born 1952. Case I94.

Severe tetraplegic choreoid and athetoid movements.

The second child of a mother age 25 at the time of delivery. The first child and the father are healthy. Father rhesus positive, homozygous and mother rhesus negative. Mother suffered from subacute nephritis. She had three attacks of nephritis during her pregnancy, with haematuria and hypertension, which necessiated hospital admission at the 5th month until term. Though repeated blood samples were taken during her pregnancy no antibodies were detected.

The child was delivered at term spontaneously after a labour of 2¾ hours. The birth weight was 8 lbs 3¼ ounces. The child appeared to be normal.

24 hours after birth however she was found to be very jaundices. The Coombs test was positive and the haemoglobin 80%. She remained jaundiced for 5 weeks and had blood transfusions on 5 occasions. For four weeks her spleen was easily palpable. For 10 days she was restless and showed slight twitching movements of the limbs at times. She fed fairly well except for the period of the twitching.

In view of the presence of jaundice and twitching she was followed up with some care in the infant clinic following discharge at the eighth week.

On three occasions up to the 18th week it was specifically stated by the clinic that limb movements and neurological examination was negative. At that age she developed an upper respirator infection with cough and she had been slightly fevered. On examination the child was miserable and fretful. Her eyes were roaming about inco-ordinately and her limbs were moved little and then stiffly, jerkily and inco-ordinately. The muscle tone was increased and on the least attempt at voluntary movement opisthotonos was suddenly assumed with extension of the limbs and supination of the forearms, flexion of the wrists and fingers. "She can't sit any more as she was doing, she's like a spring, and she suddenly goes backwards as stiff as a board."

Two weeks later evidence of respiratory infection was abscot and the child seemed well again. She was looking about her and playing with objects, rather poorly. On examination there was slight increase in the tone of the limbs and movements of the upper limbs were a little jerky but no gross abnormality was evident in the clinical findings. The sudden attacks of dystonia had disappeared.

At the age of 20 weeks she had a recurrence of her respirator infection and the attacks, exactly similar in type and severity to those previously evident returned. They persisted until the age of 30 weeks and became gradually less

severe and generalised from that time. There after they affected the upper limbs which showed sudden choreoid movements, but the trunk showed no marked involuntary movements. The legs became stiff in extension still when voluntary movement was attempted. As the attacks became less severe the mother noted a change in the fell of the limbs which gradually seemed to become stiffer than previously. The stiffness persisted. She noticed that was more marked at some times than others and that it was always exacerbated by her frequent upper respiratory infactions.

She was able to hold the head steadily from the age of 8 months, and was handling objects poorly from the 5th month but more confidently from the same time.

On examination at the age of 13 months she was alert and interested in her surroundings. She had an upper respiratory infection. She had 7 greenish teeth which were fragmenting. She had no words but attended to the voice. Did not appear to recognise her name. There was bilateral abducent paresis and nystagmus on lateral gaze of a rather coarse type. There was right facial paresis of upper motor neurone type.

The right arm was used a little for handling objects but with the fingers altogether and rather poorly. The left arm was held by the side and was used very little. On all movements athetosis of the fingers was marked in the left fingers and also evident, though not marked, in the right. As well as athetosis of the distal parts of the limbs very marked choreoid movements of the limbs were present which completely disorganised

movements of the left arm and greatly handicapped movements of the right. These were evident on the least attempt to maintain posture of the proximal parts of the limbs while the distal parts were in action. They were extremely sudden in onset and powerful, almost throwing the whole child off the mothers lap when they occured. The upper limbs tended to be place in a position of adduction at the shoulder, extension at the elbow and pronation of the forearm, and flexion of the fingers as a result of the choreoid movements.

There was a tendency to bilateral flexion associated movements of the upper limbs on voluntary activity.

The tone of the limbs was increased, very much less at total rest than when the child was even slightly alert. The tone was rigid in type. The tone was increased to a greater extend in the lower limbs than in the upper and on the left more than the right. The biceps, triceps supinator knee and ankle jerks were much increased bilaterally but especially on the left. The plantar responses were extensor on the left and flexor on the right. Sensory findings were untestable.

In their detail the attacks were rather variable in their clinical form. But they all tended to show sudden extension of the head and neck and extension of the upper limbs. In some cases only one upper limb was affected and the head was turned to one side. The legs showed no abnormality. In other cases all the limbs and the trunk were severely affected and the spasm then bore close similarities to the dystonic attacks found in cases of diplegia.

There were some differences between dyskinetic and diplegic patients even at this stage however. The impairment of consciousness seemed to be more severe in cases of dyskinesia and the attacks, usually to last longer. Many cases showed marked facial contortions and some showed deviations of the eyes and some flickering of the eye lids, finding not evident in diplegic cases.

The attacks, once they occured, usually persisted, but periods of remissions and exacerbations occured. Thus the case quoted was quite free of attacks when her first respiratory infection settled and they recurred when another respiratory infection supervened. Gradually however the attacks tended to become less generalised and the limbs on the more affected side to show movements of choreoid type, though for a period the least voluntary movement was lible to result in extension and the assumption of a position similar to that found in dystonia. The sudden increase of tone evident in the muscles when the child was showing dystonic phenomena also tended to persist. As voluntary control of the limbs improved the involuntary movements became less widespread and affected the trunk to a slighter and slighter extent, so that eventually the picture of chorea resulted in most cases without evident trunk movements.

In severely handicapped children however trunk involvement persisted and constrained extension of the limbs resulted from the involuntary choreoid movements in the limbs. None of the cases showing this pattern of involuntary movements

was associated trunk involvement could walk.

As in diplegia once voluntary control of the limbs matured flexor patterns tended to supercede extensor. It was only at this stage, when voluntary movements of the fingers developed. Therefore, the age at which athetoisis developed varied very greatly. In one case showing athetosis only, without associated choreoid movements, athetosis was evident by the age of 10 months, by which time the child was sitting along and attempting to play with her toys. In more severely affected cases however it was unusual to find athetosis before the age of 2.

The functional impairment of voluntary movement in cases of dyskinesia.

It is a matter of the greatest difficulty to detect to what degree the impairment of voluntary movement in cases of dyskinesia, is due to actual paresis of movement, except in mildly affected patients. In the latter paresis of movement is usually slight. But that some degree of paresis of movement is present is suggested by the fact that of the 17 cases in the series 12 showed abnormality of muscle tone, tendon jerks or the plantar responses, in addition to their involuntary movements. The increase of tone was slight in 4 of the seven cases in which it was observed, and it was severe in none of the cases. In three patients the increase of tone was predominantly rigid in four spastic or mixed. In three cases it was unilateral. In two it affected one arm and the opposite leg predominantly. The plantar responses were

extensor on one side in three and bilaterally in four cases. These findings are probably to be regarded as manifestations of incidental cerebral damage additional to that found in the basal ganglia and their cortical connections. In all cases in the category of dyskinesia in the series the impairment of motor function was predominantly due to the involuntary movements which occured on attempts at voluntary activity. Table IXL.

Some idea of the severity of the impairment of function is gained from an examination of what the children were able to do for themselves and to what degree the development of their motor functions were retarded. The avarage age at which unsupported walking was achieved was 2 years 5 months. Three cases aged 6 years, 9 years and 12 years were still unable to walk at the time of examination . The gaits of most of the other children were grossly abnormal. Five children over the age of 3 were unable to feed themselves completely. Only four of the seventeen cases were able to manage their buttons and laces, and thus dress themselves completely without help. Writing was extremely difficult for the majority of these children and was very clumsy and inadequate when it was possible. Three of the children over the age of 5 had speech that was so impaired that it was incomprehensible. Yet the intelligence of the children with dyskinesia was certainly not more severely impaired than in other forms of cerebral palsy.

Athetosis.

Athetosis occured as the only form of involuntary movement in only one patient who showed moderately severe

Table IXL.

Severity and extent of impairment of voluntary movement in I7 patients with dyskinesia.

Distribution.	Hemiplegic.	Triplegic.	Tetraplegic.	Total.
Severity.				
Mild.	0	2	2	4
Moderately severe.	I	I	4	6
Severe.	0	2	5	7
Total.	I	5	II	I7

tetraplegic involvement of remarkable symmetry. The involuntary movements occured only with voluntary activity in the limbs, and were more readily produced by attempts at fine movement of the fingers and hands than on movements at the proximal joints. They were produced in the arms by voluntary movements of the legs however and were slightly apparent in the opposite limb to that being used. Thus when an object was being grasped with the right hand marked involuntary extension and seperation occured in the right fingers, associated with extension of the wrist and some pronation in the forearm. At the time however associated movements, of similar type, though less marked, were evident in the fingers of the opposite hand. When she walked the feet seemed to writhe a little and the gait was rendered clumsy. At the same time some slow involuntary movements of the hands and wrists were apparent bilaterally.

There was no apparent abnormality of muscle tone and the tendon jerks and the plantar responses were normal. Associated movements of extensor type were apparent in the limbs on strenuous voluntary activity.

Choreoid movements.

Choreoid movements occured alone, without other involuntary movements in only one patient, who showed slight tetraplegic involvement. She was a mentally defective girl, 13 years old at the time of examination. There was some generalised clumsiness of voluntary movements, probably not without normal limits. In addition she showed inconstant sudden involuntary movements of the limbs, on voluntary activity, or choreoid

type. The movements were more marked when she was excited or under stress that when she was at ease. They varied in severity from day to day, and tended to be more evident towards the end of the day when she was tired than when she was fresh. The movements were more evident in the larger, proximal joints than the smaller distal parts of the limbs. This resulted in the gait being somewhat jerky as the sudden movement put the leg transiently out of alignment and the whole child became stiff momentarily. In the upper limbs the movements were more liable to occur voluntary activity necessitating more or less accurate portioning of the larger joints. Thus they occured very frequently when the child was feeding, lifting food or a cup to her lips. The limb tended to be moved suddenly and uncontrollably with considerable violence, and any object in the hand might be thrown violently out of it. The ceiling of her home was marked by the food which had been projected onto it in this manner.

Athetosis and choreoid movements in association.

Athetosis and choreoid movements occured more frequently in association than as isolated phenomena. In six patients they were the only involuntary movements which were apparent and in another six they were present in association with involuntary movement of different types.

The relative severity of the athetosis and choreoid movements in these case varied greatly, as did the extent and severity of the movements encountered. Indeed no case was very similar to any other in neurological findings.

Of the six patients showing athetoid and choreoid movements in combination without the presence of other involuntary movements, four were moderately severely and two were severely affected by the involuntary movements. Of the six patients with choreoid movements and athetosis together with other involuntary movements, one was hemiplegia, three were triplegia and two were tetraplegic. One was slightly affected, three moderately severely affected and two were severely affected.

8. The choreoid and athetoid movements were modified but not altered by the super imposition of other forms of involuntary activity, tremor, tension or dystonia. All 12 cases of choreoid and athetoid movements will be considered together therefore, and the ways in which other involuntary movements modified them will be discussed later.

9. The movements were essentially similar to the involuntary movements described in the previous two sections, but the athetosis was modified by the choreoid movements to some extent and the latter by the athetosis. In general the disturbance of function evident in the cases showing both types of movemen was more severe than in those showing the movements not in combination. This was clearly seen in a number of patients in whom athetosis predominated in some limbs and choreoid movements in others.

Female born in 1938. Case 127.

A case of moderately severe tetraplegic athetosis and choreoid movements with tension.

The third child of healthy parents. Mother was aged 36 at the time of delivery.

There was no rhesus incompatibility between the parents.

The child was born at term at home after a normal pregnancy. The labour was severe and lasted 4 hours. Delivery was spontaneous. After it the mother remember the Dr. saying that the child was stillborn and the nurse saying it was not. The birth weight was 12 lbs. Intracardic injections and mouth to mouth insufflation was applied and after 30 minutes she breathed. When the mother saw her two days later she was still black and blue all over. The left arm was bandaged. She was breast fed without apparent difficulty and no abnormality was apparent after the second week.

She smiled early but moved the limbs very poorly and sat only after the age of one year. She did not walk until the age of 2. She began to speak at the age of 3.

When the child first started to handle objects at the age of 4 she was liable to knock over anything she tried to grasp and drop anything she held.

Walking was unsteady and clumsy from the start. She seemed to flail the air with her arms and her whole body and head moved whenever she took a step. The walking became gradually less bizarre from the age of 5 when she began to attend a school for the physically handicapped. She could read by the age of 8 but could not write even her name until the age of 11. Her intelligence quotient was estimated as being 71.

On examination at the age of 14 she was a pleasant rather plain child. There was no asymmetry of the limbs. She was adolescent.

She was slow in thought and speech but answere to questions and commands were quite accurate. Her speech showed marked impairment of pronunciation of consonants and slight tendency to explosive speech, specially when excited. Her reading aloud was better than her expressive speech.

There was a right facial paresis evident at rest and on movement.

There was no restriction of voluntary movement and no contractures were evident in the limbs. The power of the limbs was fairly good but less so on the right than the left. Extension of the fingers, supination of the forearm and extension of the elbow on the right were impaired in power. There was little difference evident in the power of the legs.

All movements of the limbs were accompanied by involuntary movements. These were different in type in the different limbs. In the right arm and left leg they were athetoid in type. They affected fine and rapid movements of the distal parts of these limbs, more than the proximal. In the right arm the movements were most marked in the fingers and especially the ring and middle fingers but also involved the wrist and forearm. The movements began as a slow abduction of the fingers and thum, extension of the wrist and a marked slow undulating supination and pronation of the forearm followed. The movements persisted for several seconds at a time.

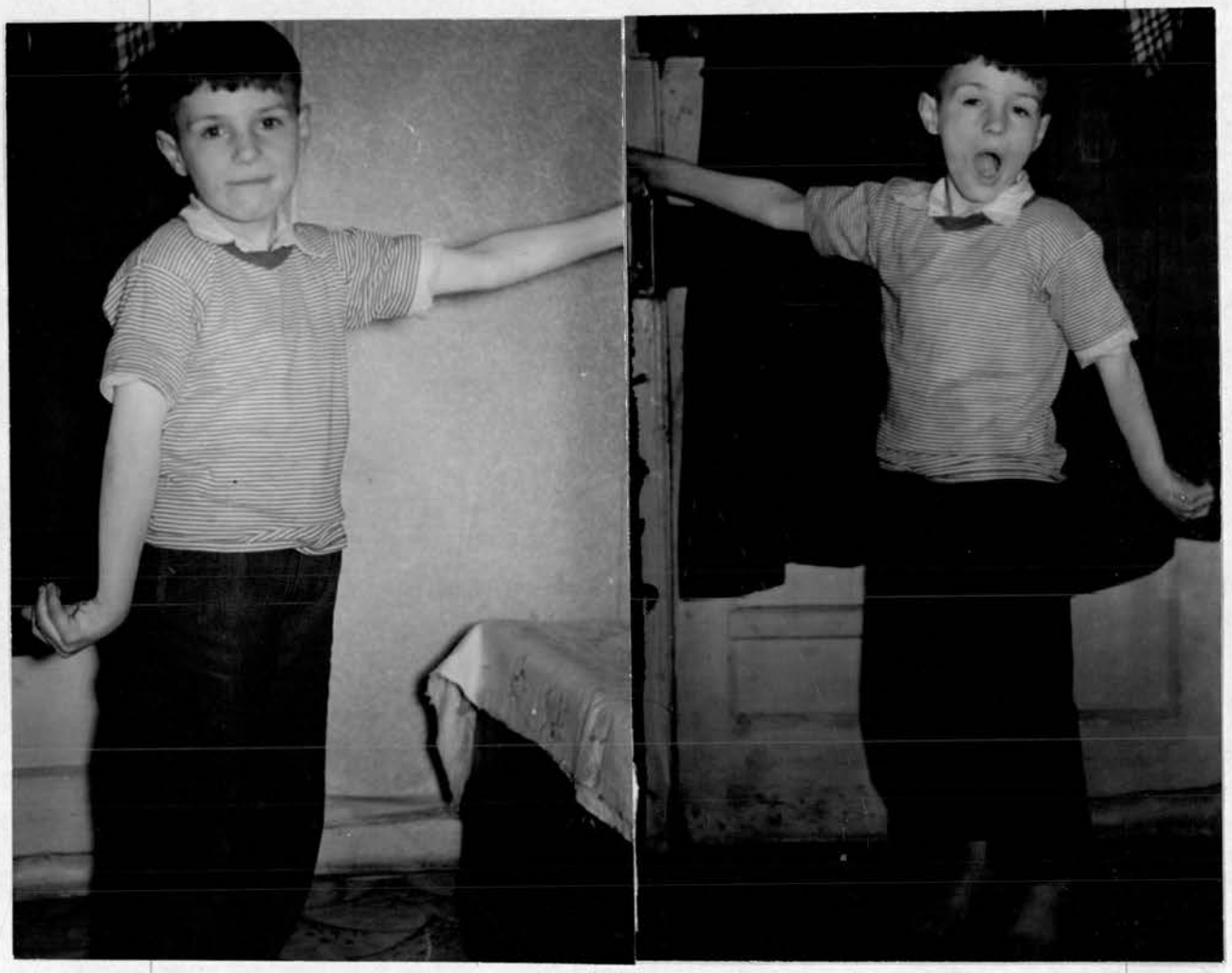
In the left foot the movements were very marked in the toes and fluctuating extension and flexion of the foot occured when voluntary movement was attempted.

Choreoid movement also accompanied all attempts at voluntary movements. These were more marked in the proximal parts of the limbs, on coarse movements than on fine movements and in the left arm and the right leg. In the left arm the movements were severe and resulted in a complete disorganisation of voluntary movements involving proximal postural control and distal manipulation. The movements were sudden, from the elbow and shoulder and resulted in a brisk extensor position of the limb being assumed with the forearm pronated and the fingers flexed. Similar movements much less severe in type occured in the right arm also. Choreoid movements in the right leg were also relatively slight.

She used the left arm, with great care for most routine activities. But by the accuracy of her knowledge of the athetoid movements in the right hand she was able to use it to a surprising degree. She used it chiefly for assisting the left and in holding objects on which she was working.

There was an increase of tone in the right arm and the left leg. This was a mixture of spasticity and rigidity in type. The tone of the other two limbs was approximately normal. The right biceps, triceps and supinator jerks were increased compared to the left. The left knee and ankle jerks were increased compared to the right. The plantar responses were bilaterally extensor.

No impairment of cutaneous sensation was apparent.



Case I97. A patient with dyskinesia, predominant movements being of choreoid type. The photograph shows the extensor associated movements present in both upper arms when the contralateral hand was grasping.

It is probably unwise to stress the differences in the characteristics of the choreoic and the athetoid movements too strongly. It is true that athetosis tended to be slow, limited to the fingers, hands, wrists, toes feet and ankles in most cases and choreoid movements to be rapid, jerky, and involve the larger joints to a greater extend than the distal parts of the limbs. On the other hand in some cases where both types of movement were present they tended to occur together and the whole limb showed involuntary movements which were typical neither of athetosis nor of choreoid movements. There is some excuse for descriptions such as "extensive and rapid athetosis" and "Slow chorea" which are found in the literature.

For the same reasons it is clearly preferable to classify these cases as showing both athetosis and choreoid movements than as being of athetoid or choreic type.

Some cases show a very marked fusing of the characteristics of both types of movement in a single limb.

The involuntary movements always tend to be worst when the involved limb is involuntary movement but the movements occur also when other limbs are moved. They may be apparent in the upper limbs when the child walks or in the contralateral arm when the least involved limb is moved. In a few cases where the chorea is severe the violence of the choreoid movement may be severe that the function of a relatively unaffected limb may be impaired by the general disturbance of posture which ensues.

Male born in 1946. Case I45.

A case showing severe tetraplegic athetosis and choreoid movements accompanied by tension.

Born to healthy parents aged 44 at the time of delivery. he was the third child, the first having died in infancy from penumonia and the second being well.

The parents show rhesus incompatability, the father being homozygous positive and the mother negative. The patients is rhesus positive.

The mother was well during pregnancy and labour occured in a nursing home at term. Delivery occured after 2 hours and was spontaneous. The child cried at onee and appeared to be normal. His weight was 7 lbs 2 oz.

He was discharged at the age of 2 weeks from the nursing home. He cried excessively in infancy and fed poorly for the first three months. He looked pale for some time but no jaundice was noted by either parent or the Dr. who was in attendance.

The first definite abnormality noted by the patient was that he could not hold up the head at the age of a year. He tended to be floppy and the head rolled about. He smiled at the age of 8 weeks and reached for objects at the age of 1 year with his right hand but not the left. He sat with support at the age of 2 and stood at the age of $3\frac{1}{2}$. He said his first words at the age of 1 but speech was incomprehensible until the age of 4.

His handling has always been clumsy and attempts at voluntary movements of the upper limbs were always accompanied

by involuntary movements. These resulted in his grasping of objects being uncertain and he dropped things frequently.

On examination he was a severely affected child with a vile temper.

There was $\frac{1}{4}$" shortening in the right arm and leg compared to the left but the trunk and head were symmetrical. Head circumference was $19\frac{1}{2}$".

He appeared to be intelligent but would not co-operate. He was distractible and energetic. He could stand with support and take a few steps unsupported but standing balance was very poor.

There was a left abducent paresis.

There was a left facial paresis only evident on movement.

The tongue showed bilateral athetosis.

The power of the limbs seemed somewhat impaired. The impairment was more marked on fine movements than on coars movements and affected extension of the fingers, wrist and elbows and supination of the forearms more markedly than shoulder movements. Dorsiflexion of the feet was weak and full extension of the knees poor. All were weaker on the left than the right.

All voluntary movements were accompanied by involuntary movements in the limbs. These were of two types.

On the left side especially, but also on the right finger movements were accompanied by athetosis, slow writhing extension of the fingers and wrist.

On movements of either upper limb marked involuntary

movements of choreoid type were evident. These were slight in the lower limbs. The movements were more evident when he attempted to maintain the position of the limb at the proximal joints than when he attempted fine movements. The involuntary movements were sudden and resulted in a marked increase of tone in the operating limb which, at the same time suddenly assumed a position of extension at the elbow, adduction at the forearm and flexion at the wrist and fingers. There was a very marked tendency to overflow of the choreoid movements so that when either upper limb was moved the opposite also showed a very marked increase of tone and a tendency to assume an extended position, and at the same time the mouth opened and the head was deviated. There was little apparent trunk involvement however.

Then he was made to move the legs voluntarily the right arm tended to assume a position of extension, the left of flexion at the elbow wrists and fingers, the forearm being pronated.

There was no generalised increase of tone apparent when the child was completely at rest but on the least movements a very marked degree of rigidity of the limbs was apparent. At rest there was some spastic increase of tone in the limbs on the left and the left biceps, triceps supinator knee and ankle jerks were much brisker on the left than the right. The right planatar response was flexor, the left extensor.

When the head was extended both upper limbs showed positions of extension, similar to that found in dystonic

attacks. The tonic neck reflex was positive on the left and negative on the right.

Supericial sensory impairment was not evident.

The degree to which involuntary movements of choreoid spread to the trunk and to the other limbs varied greatly. In severe cases of involuntary movement, head, neck, trunk and all the limbs might be involved in involuntary movements elicited by voluntary activity in a single limb. Three patients showed marked involvement of the head and neck when voluntary movement of the more affected upper limbs was attempted.

Because of the severe impairment of all forms of fine voluntary movement which results from the combination of chorea and athetosis in the limbs, only two of the 17 patients were able to manage their buttons and shoelaces unaided. The writing of all was very poor.

Of the twelve cases ten showed associated movements of the contralateral arm similar to those found in hemiplegia and diplegic paresis. In six the movements were flexor in type and in four they were extensor in type. When the children were at rest little alteration of muscle tone was encountered but the least excitement was liable to produce and increase of muscle tone and the appearance of involuntary choreoid movements. Some increase of tone, usually slight, was evident at rest in 8 patients. In three the increase of tone was slightly rigid on one side and slightly spastic on the other. In 8 the tendon jerks were exaggerated, more on one side than the other in three cases and more markedly in the legs than the arm in one case. Eight cases showed extensor

Case I73.Patient with dyskinesia who showed mixed athetoid and choreoid movements of all limbs.Much head movement from side to side when she walked.Marked athetosis of the fingers when she attempted to hold the arms outstretched.

plantar responses, 4 on only one side and 4 bilaterally.

Tremor.

Tremor of coarse type further impaired voluntary movement in two cases, one showing athetosis and in one case showing associated athetosis and choreoid movements. In the former case the tremor was evident only on voluntary movements and appeared to involve the whole of the limbs undergoing voluntary movement, proximal as well as distal joints. In the latter case also the tremor was only evident on intention and was coarse in type. It affected the distal parts of the limb the parts undergoing athetoid movement, rather than the proximal parts.

In both these cases there was quite a marked alternating increase and decrease in muscular tone in the affected limbs when the tremor was evident. The tremor was not present in limbs undergoing associated movements, the result of voluntary activity confined to other parts.

Dystonia.

Dystonic movement of the trunk and limbs was evident in three cases, one with choreoid movement and one with choreoid and athetoid movements. In all the cases the pattern of the movement was similar. Following a sudden choreoid movement of an upper limb, the limb would tend to take up a position of extension, the forearm supinated and the fingers flexed. The head and neck which had been involved to some degree in the chorea movements of the upper limb then tended to extend and the back and legs to follow suit. The opposite upper limb might or might not show extensor positions.

Synchronous with the involuntary dystonic positioning occured a very marked increase of tone in all the limbs. The dystonic position was only apparent momentarily, but was extremely powerful and resulted in a complete disorganisation of voluntary activity. None of the patients exhibiting these severe movements could walk. All were classified as being severely handicapped.

Female born in 1950. Case 37.

A case of severe tetraplegic athetoid and choreoid movements with dystonia.

Born to healthy parents. The first child. The mother was aged 24 at the time of delivery. Labour occured at term after a normal pregnancy and lasted 30 hours. Delivery was spontaneous in a nursing home. The cord had been prolapsed for 20 minutes before the child was born.

The birth weight was 7 lbs 2 oz and the child severely cayanosed, limp and apnoeic. Resuscitation, involving injections into the cord and artificial respiration resulted in breathing occuring after about 2 hours. Regular respiration was only established after 2 hours and 20 minutes. As a result of the resuscitation the child received burns of one leg from a hot water bottle.

The day after birth the child was whimpering and restless, with small range facial movements and intermittent twitching of of the limbs. His cry was high pitched. Twitching ceased after three days and thereafter apart from a marked tendency to lethargy the child appeared to be normal.

The child was eventually discharged from hospital at the age of three months by which time it was evident to the mother that the child was not normal. The limbs were stiff. The shoulders tended to be adducted, the elbow to be extended to the wrist and fingers flexed. The legs tended to be held in an extended position and the feet to be plantar flexed. She moved the limbs very little and was very still when put down. She did not attempt to use the hands until the age of one year and when she did the left was the first to be employed, the right only three months later. When the hands were first used the movements were noted to be very jerky and clumsy. She could maintain the sitting position with support from the age of 15 months. She said her fist words at the age of $2\frac{1}{2}$, and about the same age developed a great interest in picture books and passing traffic.

True involuntary movements of the limbs became apparent at the age of about 6 months. They resulted in the child suddenly becoming stiff the back arched and the neck hyperextended. The upper limbs showed shoulder adducted, the elbows extended, the forearms pronated and the wrists and fingers flexed. The legs strongly extended and the feet plantar flexed. This movement resulted from any sudden change in the position of the child or attempt at voluntary movement of the limbs. This persisted until the age of 3 but by this time the involvement of the trunk was less marked.

Twitching attacks of the limbs with impairment of conciousness which lasted for a few seconds became apparent at the age of 6 months at the same time as attacks of dystonia

became evident. These attacks of impairment of consciousness began with twitching of the eyes of flickering type and facial movements. The fingers worked bilaterally and the limbs became stiff, the back arched. During the time the back was arched for perhaps ½ a second the childs conciousness appeared to be severely impaired. The arching of the back and the stiffness of the limbs relaxed suddenly and was succeeded by a period in which the child felt flabby and conciousness was fully regained. Initially the attacks occured 2 - 3 times a day, but at time when she was suffering from one of her periodic respiratory infections they were very much more frequent at the age of 2½ and were only occasional from the age of 3.

She had always drooled.

Examination at the age of three year showed a severly affected child.

She comprehended most of what was said to her and enjoyed her surroundings. She had six words. Her consanants were very poorly pronounced and explosive speech was evident.

She was able to sit with support and momentarily without it. No standing balance.

There was bilateral abducent paresis with marked swinging nystagmus bilaterally when she attempted to fix the gaze. Involuntary movements of the face bilaterally were present when she attempted to speak.

Swallowing was infrequent and drooling was present.

The power of the limbs appeared to be normal and there was no restriction of voluntary movement because of contracture.

She moved the left arm more than the right. Her hand hand movements were of infantile type. They were accompanied bilaterally by marked slow writhing athetosis of the fingers wrists and forearms. The tone of the limbs was increased moderately more in the right limbs than the left. It was rigid in type. The tendon jerks were increased in all limbs. The plantar responses were equivocal.

Voluntary movements of the proximal limbs was accompanied by movements of choreoid type. These were severe and affected the right arm and leg more severely than the left. The choreoid movements occured suddenly and were extremely powerful. They resulted in a generalised extension of both upper limbs associated with hyperextension of the neck and trunk, so that a modified dystonic position was assumed. This movement occured whenever much voluntary movement of either upper limb was attempted but was more easily reproduced from the right than the left. The movement was sufficiently severe to throw the child backwards when sitting with support and disintegrate any attempt at voluntary activity of constructive nature.

During the examination two attacks with marked impairment of conciousness occured similar to those discribed by the mother.

These began with twitching of the eyelids and marked swinging nystagmus. Twitching movements of the face and fingers followed and then dystonic movement ensued. The duration of the attacks was 8 0 seconds in one and 9 0 seconds in another.

Sensory findings were untestable.

It is interesting that two of the three patients with dystonic movements showed marked associated movements of the extensor type on voluntary activity of the limbs. Clearly there is some similarity between these cases and those in the dystonic stage of diplegia.

Tension.

Sudden generalised increase of muscular tension affecting equally agonists and antagonists was almost invariable in those showing choreoid movements when voluntary activity was attempted. The degree to which this tension affected limbs other than the one being move was variable.

In four cases, one with choreoid movements only and three with athetoid movements also, the tension was severe. In these patients voluntary movements were checked and sometimes rendered impotent by its occurence, as in those with dystonia.

Other causes of marked increase in muscular tone in these cases were conditions of excitement, fatigue or mental stress. As with choreoid movements tension was found to vary very greatly in its severity in the same patients at different times and in different conditions. Cases seen briefly at clinic examinations had, in fact frequently been diagnosed as cases of generalised rigidity whereas careful examination with the child at ease would have shown that, in fact, muscle tone at rest was normal.

It was interesting to observe that patients with marked increase of tone voluntary movement function was sometimes quite as much impaired by this as it was by actual choreoid movement of the limb. It seems much more likely that the

increase of tone is a true reflex manifestation than that it is a concious act on the part of the child to lessen his involuntary movements as has been suggested. Phelps.1943.

In one patient the tension on voluntary movement was evident in three limbs, and one arm was much less affected by it. In other three patients all the limbs showed the increase of tone though to different degrees. In two of the patients the state of tension was also found in the contralateral upper limb to that being used. In both extension of contralateral limb was evident.

Case 145 as quoted on page.

It seems probable that the condition of involuntary tension is somewhat similar in origin to that which accompanies involuntary movements of the dystonic type.

Other findings in the limbs.

Vasomotor changes similar to those found in hemiplegic cases were encountered in none of the patients classified as suffering from dyskinesia. The feet of some of the more severely incapacitated patients confined to bed or wheel chairs were found to be colder than the hands. Cyanosis was encountered in none of the cases.

Generalised dwarfing was not a feature of dyskinesia and the heights of children over the age of 5 appeared to be within the range of normal according to available figures. Asymmetry of the limbs was not marked. In only one case was one upper limb more than $\frac{1}{2}$" shorter than the opposite upper

limb, and in two cases shortening of 1/3" was found, with less marked dwarfing in the corresponding lower limbs, in all three cases.

In no case was disturbance of superficial sensation or joint sens noted. In view of the cases of hemichorea and hemiathetosis with hemi anaesthesia recorded in the early literature this was something of a disappointment. Charcot.1868.

Speech defects in dyskinesia.

In addition to the speech defects encountered in diplegic hemiplegic patients, which were also found in those suffering from dyskinesia, another type of speech disorder was found in the latter group. Owing to poor co-ordination of voluntary respiratory activity the speech was ill controlled in volume and tended to be checked or forced by involuntary movements of the respiratory muscles. This phenomenon of explosive speech was very marked in four of those with some speech and seemed to be a major cause of failure to develop speech in three of the cases in whom no comprehensible speech was present.

The types of speech disorder encountered are shown in table XXXI. Dyslalia alone was the commonest disorder and occured in six patients, one hemiplegic in distribution, three triplegic and two tetraplegic. Two patients were mildly affected by dyskinesia, three moderately severely affected and one severely affected. Explosive and dyslalic speech was found in three patients, two moderately severely and one severely affected by dyskinesia. Stutter with dyslalia was noted in one patient.

Table XXXVI.

Speech defects in 17 patients with dyskinesia.

Type of defect.	Number.
Explosive speech with dyslalia.	3
Explosive speech with dyslalia and hesitation.	I
Dyslalia and stutter.	I
Dyslalia only.	6
Rudimentary speech only.	4
Total speech defects.	15.

Four patients showed no speech or their attempts at speech were so poor as to incomprehensible. All but one child, who was age 15 months at the time of examination were over the age of two and were severe cases of dyskinesia.

Thus in all 15 of the 17 cases of dyskinesia showed speech defect sufficiently severe in degree to impair the comprehension of what they attempted to say.

Cranial nerve involvement in dyskinesia.

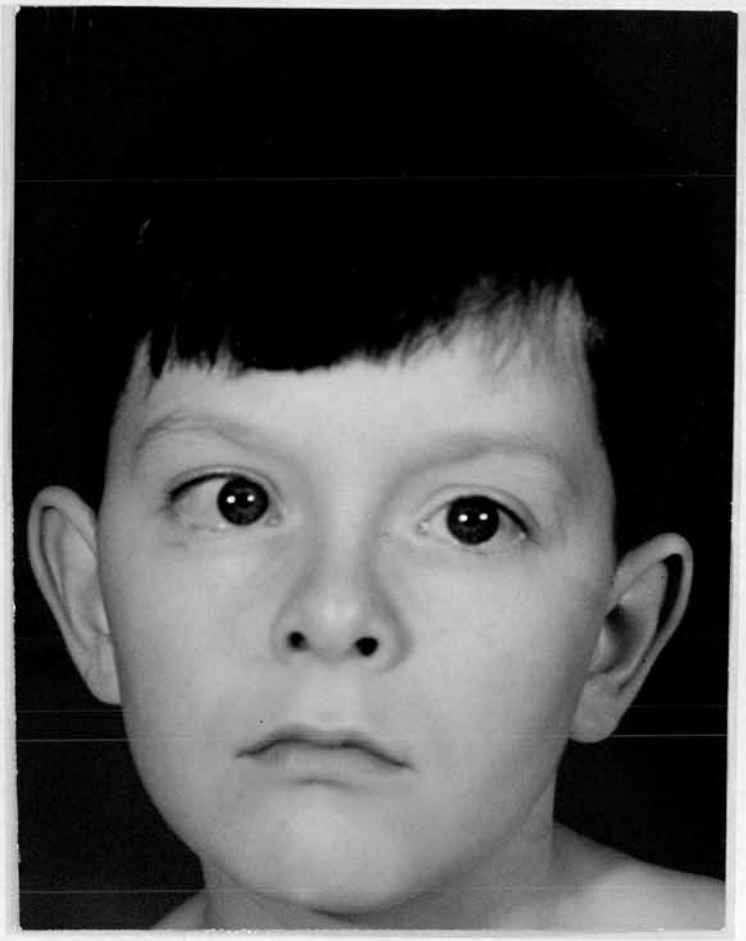
Visual impairment.

Impairment of vision due to optic atrophy or field defects was not found in any of the patients with dyskinesia. One patientsshowed severe myopia and another severe astigmatic refractive error. Two cases showed inequality of the pupils, though these reacted normally to light and accomodation.

Eye movements.

Strabismus was found in six cases. It was due to unilateral abducent weakness in two patients and to bilateral abducent weakness in four.

Nystagmus was evident in five cases. In all it was most marked when an attempt was made to fix the gaze and two of the children, who were able to read held the head and eyes in curious positions which they assumed in order to minimise the effect of the nystagmus. The eyes were held to the side to which the nystagmus was least severe and the head rotated to the opposite side. In three of those with nystagmus strabismus at rest was apparent, in one unilateral abducent paresis and in two bilateral abducent paresis.



Case II2. Strabismus due to right abducent weakness in a case with mixed choreoid and athetoid movements.

Facial involvement.

Facial paresis of upper motor neurone type was found in five patients. Only in one was facial asymmetry present at rest. In the other four patients it was only manifest on movement. Facial overaction was encountered in one or these patients. In the other three there was facial lag on the affected side on emotional and voluntary movements.

Involuntary movements of the face, fluctuant or tremulous in type, occured on voluntary facial movement in six patients. In five of these the involuntary movements were unilateral; in one they were bilateral. They were evident in all on speech, or attempts at speech as well as on movements elicited for the purposes of examination. In all these patients overflow of involuntary movement from the upper limb on the affected side to the face occured. In some patients the involuntary facial movement was marked and associated with opening of the mouth and very bizarre facial expressions.

Hearing.

High frequency deafness was confirmed by audiometry in seven of the II cases of dyskinesia tested. The audiograms of 4 cases were normal. In six cases audiometry had not been performed.

High frequency deafness has been noted to occur frequently in cases of dyskinesia and is considered to be an important cause of speech defect, as the child is unable to hear speech normally because of it. Asher. 1952.

Swallowing.

In four patients, all of whom showed dyskinesia of

moderately severe or severe degree swallowing was impaired, and drooling was marked to the age of 5 or 6. In two of these patients swallowing occured less frequently than once in two minutes on the average. All the children who showed difficulty in swallowing showed speech defects. Two had no comprehensible speech, and the two others showed explosive speech and dyslalia. Three of the four patients with impairment of swallowing had facial involvement. Two had involvement of the tongue.

Impairment of voluntary movements of the tongue.

Abnormality of the movements of the tongue was noted in seven cases. In all these cases voluntary movements of the tongue were impaired by involuntary movements, usually of a fluctuating character. Because of them the full range of voluntary movements was impossible and the child was unable to maintain the tongue even momentarily in any one position.

All those who showed tongue involvement also showed speech defects. In two speech was so poor as to be incomprehensible. In three there was dyslalia and explosive speech and in two dyslalia only.

Intellectual impairment in dyskinesia.

The schooling received by the 17 cases of dyskinesia and their intelligence as measured by different observers in different circumstances and by different tests are shown in tables XXXVII & XXXVIII. It will be noted that of the 5 children with intelligent quotients of over 85 only two attended normal school. This indicates the severity of the educational handicap suffered by children with dyskinesia. It is

Table XXXVII

The intelligence quotients of 17 patients with dyskinesia.

Intelligence quotients.	115-	110-	85-	70-	55-	Under 55.	Untested.	Total
Number of patients.	1	2	2	4	2	1	5	17.

Table XXXVIII

The schooling of 17 patients with dyskinesia.

School.	Number of patients.
Normal schools.	2
Schools for the physically handicapped.	7
Schools for the mentally handicapped.	2
Ineducable.	1
Not yet at school.	5
Total.	17.

interesting that the numbers of cases of normal intelligence who find their way to special schoolf for those with cerebral palsy include a high proportion of children with dyskinesia. The severity of the educational disability suffered by these cases is out of proportion to their intellectual impairment.

The numbers of cases in this survey are insufficient for any opinion to be given as to the correctness of the impression that children with dyskinesia show less impairment of intelligence than do other types of cerebral palsy. Dunsdon,1952; Evans, 1948.

Overactivity.

One intelligent child with triplegic dyskinesia of moderate severity showed overactive behaviour similar to that described when hemiplegic cases were discussed.

Epilepsy in patients with dyskinesia.

Four patients showed epileptic phenomena. One patient showed petit mal beginning at the age of 6 months and continuing at the age of $2\frac{1}{2}$ at the time of examination, 15 to 20 times a day. Three patients showed fits of other types. (One of these, case 37 has been noted on page).

There was similarity to the dystonic attacks noted in deplegic patients in this except that both the movements and the impairment of conciousness were evident for a longer time than typically found in the diplegic patients. The attacks are also rather similar to the so called striatal attacks found in some cases of enceplatis lethargica.

One patient showed attacks lasting one minute to 80 seconds and occuring 15 - 20 times a day. In these conciousness was impaired but not lost. The attacks began with a sudden incoordinate movement of the head and neck, the eyes showed marked nystagmus and the left shoulder was moved rapidly and incoordinately from the trapezius. No abnormal movements of the legs were noted. After about 1 minute the attacks ceased and she seemed normal again. During the attacks the tone of the upper limbs was very greatly increased. The girl was a severe tetraplegic case. The attacks began at the age of 6 months and were still present at the age of 3.

One severely tetraplegic girl aged 6 started to have attacks at the age of 1 year and these were evident at the time of examination. Her attacks began with sudden severe impairment, but not absolute loss of conciousness, and a marked rigidity appeared in the limbs and trunk. The head and neck extended and deviated to the right. The upper limbs flexed briskly and sometimes hit her severely in the face. The lower limbs extended. After about 1 minute the child seemed normal. Sometimes she was incontinent during the attack.

It is probably of significance that in these three patients the first of these a typical epileptic attack began when the child was beginning to hold up the head. Initially there was a story of quite marked extension of the head, neck and limbs in all three, but as the attacks continued they altered in form and were associated with flexor positions of the limbs to a greater extent. The attacks were becoming

less frequent in two of the cases in whom postural activity was improving, but the first case quoted, (case 178) where no improvement had been noted they persisted.

Other findings in cases with dyskinesia.

One patient showed congenital cleft palate and hare lip. One patient showed bilateral congenital dislocation of the hips which it had not been possible to correct by orthopaedic means. One patient was stated to have shown "club feet and hands" immediately after birth, by the hospital notes. The feet had been energetically treated from the age of 2 days and at the age of 10, when she was examined only slight talipes equino-varus deformity was evident bilaterally.

One patient, with kernicterus showed decaying green teeth. Five of the seven patients still with their first dentition showed very decayed teeth. A history of very poor teeth in the first dentition was obtained in 6 of the other ten cases. The finding of poor teeth did not seem to be related to the dyskinesia being due to rhesus incompatability or not.

Chapter 6.

The clinical findings in patients with bilateral hemiplegia.

Eight patients one male and seven females with cerebral palsy were classified as suffering from bilateral hemiplegia. In all the impairment of motor function was paretic in type and more severe in the upper limbs than the lower. In all contractures were evident and all but one, who was considered to showemoderately severe impairment, showed severe impairment of voluntary movement. All but one of the patients were bedridden.

All the patients were considered ineducable, either by the school authorities or the examiner. In none of the patients tested was the intelligence quotient more than 50. All the patients showed cranial nerve involvement and swallowing was difficult or impossible in six. Only one child showed any comprehensible speech. All but two of the patients were microcephalic, having a head circumference of less than 19" after the age of 4. The patients are similar to that described by Freud, and diffenciated by him, from diplegic disorders. Freud. 1897.

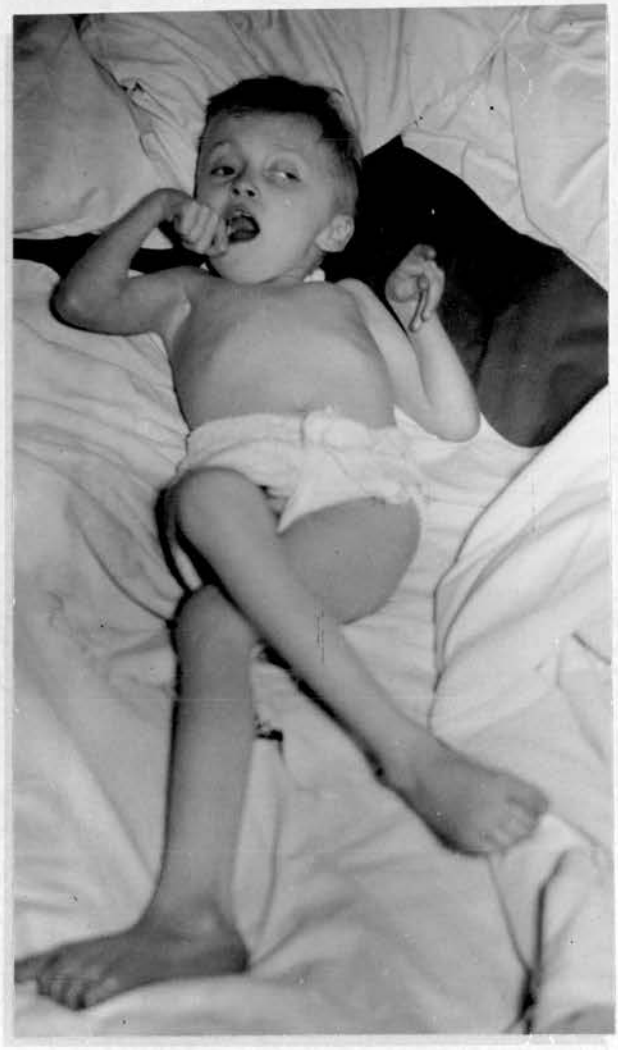
Motor impairment.

In all the patients with double hemiplegia the voluntary control of the limbs was deficient. It was difficult to gauge the severity of the deficiency because of the severe intellectual impairment, disuse contractures and lack of cooperation shown by all the patients.

Only one of the patients was not classified as suffering from severe impairment of voluntary control of the limbs.

She was a grossly overactive girl of 6, who was considered to be moderately severely affected. She was capable of walking unsupported, though her gait was very spastic and unsteady. She was able to grasp with both upper limbs but did so in an infantile fashion, using all the fingers, not the thumb and forefinger, of the hands. Her handling was grossly clumbsy. Release of the grasp was very poorly co-ordinated and objects were thrown from the hands rather than released from the grasp. Both hands were equally affected and marked athetosis of the fingers was evident. Full extension of both wrists and elbows and supination of the forearms was limited by contracture. The feet were incapable of passive dorsiflexion to the right angle owing to contracture and the hips showed some degree of flexion deformity. Associated movements of the upper limbs were marked and a flexion type when she walked. There was generalised spastic increase of tone more marked in the arms than the legs and rather more severe on the right side than the left, with marked bilateral exaggeration of the tendon jerks.

Of the seven patients classified as suffering from bilateral hemiplegia of severe degree all were bedridden and showed severe generalised flexion contractures of the limbs. None was able to feed himself. All were incontinent of urine and faeces. The degree of voluntary movement present in the limbs varied but was poor in all the patients. Only one seemed to take the least interest in attempting to handle objects. He was able to grasp them but not to release them



Case of bilateral hemiplegia who was totally bedridden on account of severe generalised contractures.

from the grasp. Some of the other patients were able to grasp objects placed in their hands but inall the grasp was achieved by using all the fingers and not the thumb and fore finger. None was able to stand or to sit without support. Only one was able to sit even with support. Only three were able to hold up the head.

In all there was marked increase of tone in the limbs, more severe in the arms than the legs and predominantly spastic in type, though some rigidity, the result of contracture, was also evident. The tendon jerks were increased markedly in all. The degree of exaggeration of the tendon jerks was usually more marked on one side than the other. The side showing the greater exaggeration was not necessarily the more severely affected side, for in some contracture had limited the response of the muscle to the stimulation.

In all the plantars were extensor, though whether the planatar response was of the infantile withdrawal type of a true Babinski response was sometimes difficult to determine.

Female born 1950. Case 6.

A case of severe bilateral hemiplegia.

The only illegitmate child of a mother aged 18 at the time of delivery. She was well during pregnancy except for mild preeclamptic toxaemia in the later months. The child was delivered 2 weeks after term in hospital spontaneously after a labour of 6½ hours.

The weight was 7 lbs 7 oz. The child cried at once. She was extremely lethargic and feeding gave rise to great

difficulty in the early days. She was floppy and went to sleep at the breast. She would take milk into the mouth but would not swallow it and sometimes would cough it back. She remained lathargic following discharge from hospital and feeding difficulty persisted. She was admitted to hospital with broncho pneumonia at the age of 6 weeks and again at three months.

At the age of 6 months the mother was doubtful if she could see and noted that the limbs were becoming increasingly bent and stiff. She made no movement when laid down and would be found in the same position hours later. She appeared not to recognise people, even her mother and showed no interest in her surroundings. She did not smile.

She began to hold the head up at the age of $2\frac{1}{2}$ but could not sit without support. She had smiled shortly before this. She appeared to recognise the mother and gurgled with pleasure and cried when uncomfortable. She made no effort to handle objects.

Examination at the age of 3. Height 33". Head circumference $17\frac{5}{8}$". The fontanelle was one finger open still. She had a mass of fine black hair which extended onto the trapzius muscles posteriorly. There was marked hyperteleorism.

She showed little interest in her surroundings but looked up transiently as people came and went. She could hear. She had no speech. She spend her time playing with her long hyperextensible fingers without any obvious intention. She could sit with support.

There was a left abducent paresis.

A right facial paresis was present.

Swallowing was infrequent, performed with an obvious effort and drooling was marked.

Marked flexion contractures of all the limbs were present. And no voluntary movements of the forearms, elbows or shoulders were present. She was able to play with her fingers however but could not be induced to hold anything in them. There was slight voluntary movement at the knees, which could be extended a few degrees. No dorsiflexion of the feet was possible. Bilateral equino-varus deformity was present.

The jerks were impossible to elicit owing to contracture. The plantars were extensor, but stimulation of the sole of the foot was accompanied by a writhing withdrawal reaction of the whole limb stimulated.

The sensory findings were untestable.

Athetosis of the fingers, similar to that found in cases of unilateral hemiplegia was evident on attempted movement of the limbs in four of the cases of bilateral hemiplegia.

Dwarfing of the limbs.

The limbs, and especially the upper limbs appeared to be small and generally poorly developed with reduction of girth, as well as of length, in all the patients but because of the severe contractures in most, it was difficult to obtain accurate measurements. In only two cases was marked asymmetry of the limbs apparent. One patient showed apparent shortening of $1\frac{1}{2}$" in the upper limb and $\frac{3}{4}$" in the lower limb on one side compared to the opposite.

Another patient showed I" apparent shortening in the one upper limb and ½" shortening in the lower compared to those on the opposite side. It was interesting to observe, in both these cases that the degree of contracture was approximately equally severe bilaterally.

Vasomotor changed in the limbs.

As in cases of unilateral hemiplegia vasomotor changes were evident in the limbs of those showing bilateral hemiplegia. In all cases the extremities were cold and the hands more severely affected than the feet. In three cases persistent cyanosis of the hands and feet was evident. In one of these cases there were marked chilblains of the hands and feet and a varicose ulcer was present over one tibia. This was similar in type to varicose ulcers of the adult.

Retardation of growth.

Unfortunately the number of cases is too small for the degree of growth impairment to be considered in a statistical manner and since the time of onset of the bilateral hemiplegia was uncertain in some patients this would have been a matter of great difficulty, had it been possible.

All the patients were smaller than would have been expected if they had been normal children, however, and the apparent dwarfing appeared to be more marked in older, than younger patients.

The head circumference in patients with bilateral hemiplegia.

The front occipital circumference was measured in all cases with bilateral hemiplegia and was found to be reduced,

according to the usually quoted figures, in all but two. All patients except these two had head circumferences below 19". In 5 the head circumference was $18\frac{1}{2}$" or less and in two less than 18". None of the patients were less than $3\frac{1}{2}$ years at the time of measurement.

The shape of the heads varied greatly. Most cases showed a severe degree of occipital flattening.

One patient showed a head circumference which was in the lower part of the normal range. One patient, who had had cerebral thrombophlebitis complicated by meningitis in infancy showed arrested hydrocephalus, and a head circumference of $21\frac{3}{4}$" at the age of eleven.

Intellectual impairment of cases with bilateral hemiplegia.

All the five patients over the age of 5 who were suffering from bilateral hemiplegia had been classed as ineducable by the education authorities. Of the three under the age of 5 all were grossly defective.

Only one patient had any comprehensible speech, and he could only imitate a few words which he had heard often repeated by his relatives or on the wireless. None of the other patients had any comprehensible speech and none was able to express his wants even in sign language. How much the patients appreciated of their environment was impossible to estimate. It was clearly very little.

Owing to the absolute lack of volition of most cases, contracture of even slightly affected limbs was inevitable.

The one patient who had independent walking showed grossly overactive and disordered behaviour.

She was never still for a moment, dashed clumsily about, falling frequently and damaging herself and all accessible furniture in her course. Her behaviour was similar in type to that described when unilateral hemiplegia was discussed. Her behaviour could only be described as animal in type.

Visual impairment.

Vision was defective to some extent in all the cases of bilateral hemiplegia. Only two patients, those with hydrocephalus and limited speech, and with independent walking and gross behaviour disturbance respectively, showed any ability to detect more than the difference between light and darkness. The over-active patient appeared to have hemianopia and to have grossly impaired visual perception. She showed bilateral coarse nystagmus. The hydrocephalic patient was able to recognise doors opening and shutting and to follow the movements of people about his room.

One patient showed severe unilater cataract. Four of the patients showed well marked bilateral optic atrophy.

Cranial nerve involvement.

All the patients with bilateral hemiplegia showed evidence of cranial nerve involvement.

Strabismus.

Six patients showed strabismus. In three there was unilateral abducent paresis. In one bilateral abducent was present. Two patients showed bilateral third nerve paresis, one with associated unilateral abducent paresis.

Facial involvement.

Facial paresis was difficult to examine on account of the inability of the children to co-operate. Four patients appeared to have more or less symmetrical bilateral facial paresis of upper motor neurone type. Three patients appeared to have bilateral facial paresis more marked on one side than the other. One patient, only, appeared to have paresis confined to one side of the face. Thus all the eight cases showed either unilateral or bilateral facial paresis.

Hearing.

This was quite impossible to test in most cases as it was impossible to distinguish lack of comprehension from deafness and appreciation of vibration from actual hearing.

Swallowing.

In all cases a history of difficulty in swallowing and of drooling was elicited. Difficulty in swallowing was still evident at the time of examination in all but two patients. None of these were able to take normal solid food and slops comprised their diet. Drooling was evident at the time of examination in four of the six patients with difficulty swallowing.

Owing to impairment of swallowing much aspiration of food occured in most cases. Five of the eight patients with bilateral hemiplegia had a history of hospital admissions for respiratory infections on one or more occasions. It is probable that aspiration of food material was the most frequent cause of these. The patience of the mothers in the feeding of

these hopelessly defective children was quite unbelievably good.

The tongue.

It was impossible to test tongue movements in the patients with bilateral hemiplegia.

Epilepsy.

Grand mal attacks were present in five patients. In one of the patients grand mal attacks with loss of conciousness for up to 30 minutes occured two to four times a day. One patient had attacks two or three times a week with impairment of conciousness lasting for about 5 minutes on each occasion. In three patients the grand mal attacks occured at intervals of several weeks or months. In all these patients the effect of antiepeileptic drugs was disappointing. One patient showed more or less continual twitching of the face and arms, which only ceased on the administration of very large quantities of sedatives.

The level of conciousness of the patients with bilateral hemiplegia appeared to vary from minute to minute and from day to day to a degree which was difficult to assess.

Associated abnormalities.

Three patients, all female and showing microcephaly had a curious hair line posteriorly. It extended low down on the neck and tended to spread slightly over the trapezius muscles bilaterally. The hair was black and bushy, fine and curly in all three cases. Two of these patients showed excessive hair over the extensor surfaces or the upper limbs.

One patient showed moderately severe talipes equino-varus deformities of both feet which had been present from birth. One patient showed multiple skeletal deformities. Talipes equino-varus was present in one foot and valgus deformity of the other. There was a marked lumbar scoliosis with three hemivertebrae and bilateral congenital dislocations of the hips. One patient had a congenital heart lesion, thought to be interventricular septal defect.

One patient showed marked evidence of healed rickets.

Clinical findings in other cases of cerebral palsy.

Two patients could not properly be classified in any of the previous categories.

One of these was a moderately severely handicapped girl with severe arthrogryposis. She also had bilateral third nerve paresis, bilateral facial paresis of upper motor neurone type and was unable voluntarily to shut the eyes. Involuntary shutting of the eyes was normal. Swallowing shoulder and tongue movements were normal. Her speech was normal. She attended a school for physically handicapped children and her intelligence quotient had been estimated as being 62. She was considered to be a case of cerebral palsy confined to the cranial nerves.

The other patient was a case of post-encephalitic parkinsonism. He had been acutely ill three years before examination and following his illness his thought became slow, his limb rigid and he developed a generalised tremor. He swallowed infrequently and drooled. His speech becam slow

and very slurred so that it was hardly comprehensible. All his movements were slow and poorly controlled. His gait was festinant. He was able to walk only short distances without support and is taught at home by a teacher visiting the physically handicapped. His intelligence quotient was 135. He was considered to be severely affected.

SECTION 3

CHAPTER 1

The incidence of cerebral palsy amongst children in Edinburgh.

As a result of the survey it is possible to present some estimate of the incidence of cerebral palsy in the City of Edinburgh, for children born between the years 1938 and 1952 inclusive. Of children born during this period 208 were found to suffer from cerebral palsy. The estimated population of this age group in Edinburgh derived from the figures of the Education Department and the Registrar General for Scotland is 104,285. The incidence of cerebral palsy in children, aged up to 15 years inclusive, is 1.991 per 1,000.

The incidence of cerebral palsy by year of birth.

In table XL. are shown the incidence figures for children born in three and five year period from 1938 to 1952. Some explanation for the apparent variations in incidence are necessary.

It will be observed that for the two 3-year periods from 1944 to 1946 and from 1947 to 1949 the incidence of cerebral palsy was approximately 2.5 cases per 1,000 of the population. The incidence in both younger and older children is lower. It is necessary to determine whether the differences in incidence at different ages are real or apparent.

The /

TABLE XL.

The population of Edinburgh by year of birgh, 1938-52, based on figures obtained from the Education Authority and estimates of the Registrar General. The incidence of cerebral palsy by year of birth.

	Numbers of patients				Incidence per 1000 in 3 year periods.		
Year of birth.	Population	Hemiplegia	Diplegia	All types	Hemiplegia	Diplegia	All types
1938	6749	10	3	17			
1939	6574	3	8	13	.819	.563	1.945
1940	6209	3	0	8			
1941	6099	4	2	10			
1942	6388	4	4	13	.631	.578	1.839
1943	6538	4	5	12			
1944	6533	7	11	24			
1945	5993	4	3	9	.930	1.028	2.546
1946	7902	8	7	19			
1947	8900	8	7	21			
1948	7700	6	4	14	.871	.996	2.448
1949	7500	7	13	24			
1950	7200	5	3	11			
1951	7100	1	6	8	.330	.566	1.132
1952	6900	1	3	5			
1938-52	104285	75	79	208	0.719	0.757	1.991

The incidence of cerebral palsy amongst children born in 1950 to 1952.

The incidence of cerebral palsy in children during the first three years of life was smaller, 1.123 cases per 1,000, than that for older children. The numbers of cases gradually increased in the first four years. The reasons for the lower incidence in younger children are multiple and complicated. Clearly it could be due either to defective ascertainment of patients with cerebral palsy, or it could be due to there being fewer patients suffering from the condition in the younger age groups. The lower incidence may be apparent or it may be real.

The proportion of patients developing cerebral palsy after the age of three is small. Only seven patients with hemiplegia, three with ataxia, one with bilateral hemiplegia and one with postencephalitic Parkinsonism who developed their palsy after the age of three were encountered and none with diplegia. Of the 208 patients in the series only 12 or approximately 5.8% developed cerebral palsy after the age of three.

Therefore, the lower incidence of cerebral palsy in younger children must be assumed, for the most part, to be due to defective ascertainment. Ascertainment was defective /

Table XLI.

The age of the occurence of the first abnormality, the correct diagnosis, first visit to Doctor and first clinic referral in 79 patients with diplegia.

Age.	First abnormality.	First visit to doctor.	First clinic referral.	Correct diagnosis.
Under 6 months.	3I	23	8	3
Over 6 months under one year.	23	I0	I2	I
Over one year under I8 months.	I0	25	I2	I4
Over I8 months under 2 years.	II	I2	I2	27
Over 2 years under 4 years.	2	7	26	I9
Over 4 years.	0	0	8	I4
Unknown.	2	2	I	I
Totals.	79	79	79	79

defective for a number of reasons. In the first place it was impossible to survey such large groups of children under the age of four as it was of older children who were at school. The ascertainment had to be carried out almost entirely at second hand on the basis of cases who had been reported to hospitals, and infant clinics, or who were known to general practitioners. Because there was considerable delay in cases being correctly diagnosed by general practitioners and further delay in their referral for treatment at clinics, cases did not become known to the survey until a considerable lapse of time after the appearance of the first manifestations of palsy.

Thus delay in diagnosis and delay in the child reaching hospital clinics were the predominant reasons for the apparently lower incidence of cerebral palsy in the first three years of life. This was well shown by the 79 patients suffering from diplegia in the series. Only one diplegic patient appeared to have acquired his disorder as a result of factors operating later than the newborn period; the remaining 78 cases apparently developed the condition as a result of abnormalities occurring in the prenatal, natal or neonatal periods. It will be seen from table XII. that the majority of these patients /

patients had shown symptoms or signs of diplegia before the age of one year, and that most were taken to their doctors shortly after the first manifestations of cerebral palsy had occurred. Yet there was an average delay of over one year in reaching the diagnosis. In no less than 14 cases diplegia was only diagnosed at the age of 4 or later.

It has been emphasised that the early stages of diplegia are little understood and are largely neglected in standard textbooks, in which invariably cases are described in the late stage of contracture. Nevertheless the early manifestations are striking and should have certainly led to full neurological examination in the majority of the patients at an early age. The first abnormalities noted by the mothers in the cases of diplegia are shown in table XLII. In spite of these early manifestations of neurological abnormality most patients reach treatment only when contractures are well in evidence, one or two or more years after they have occurred. In present circumstances, therefore, it is inevitable that a falsely low figure for the incidence of diplegia should be obtained in the first three years of life, unless large scale sampling is undertaken.

It is thought that the lower incidence of cerebral palsy amongst children born between 1950 and 1952 found during /

Table XLII.

The nature of the first neurological abnormality noted by the mother or guardian in 79 children with diplegia.

Nature of the abnormality.	Number of patients.
Paresis, rigidity or extensor spasms.	45.
Epilepsy.	14.
Blindness.	5.
Mental retardation.	8.
Behaviour disorder.	2.
Other abnormality.	3.
Unknown.	2.
Total.	79.

during the survey is chiefly due to delay in recognising the nature of the early manifestations of the disorder. It cannot be regarded as being the result of there being any significantly smaller number of patients with cerebral palsy in the younger age group.

The incidence of cerebral palsy in children born between 1938 and 1943 compared to that in those born between 1944 and 1949.

It will be observed that there is apparently a lower incidence of cerebral palsy amongst children born between 1938 and 1943 than amongst those born between 1944 and 1949. In the older children the incidence is approximately 1.9 cases per 1,000, in the younger approximately 2.5 per 1,000. Table XL.

It is necessary to consider whether the lower incidence quoted for children born between 1938 and 1943 is the result of more defective ascertainment, than amongst children born between 1944 and 1949. Because access to the older children was easier than to the younger as all the former were at school or known to school medical authorities and some of the latter were under school age, inspection of the children born between 1938 and 1943 was fuller than for those born between 1944 and 1949. It is true that children over the age of 12 had ceased to attend children's hospitals but access to patients in orthopaedic and neurological clinics and wards /

wards in the general hospitals was very readily granted. Moreover the notes of possible cases of cerebral palsy in children's hospitals for the last 15 years were inspected. It seems unlikely therefore that many patients with the condition in the older age group were missed. It is unlikely that the apparent difference in incidence between children born in 1938 to 1943 and 1944 to 1949 is wholly due to defective ascertainment of cases in the former age group.

The possible reasons for there being a smaller incidence of cerebral palsy amongst children born between 1938 and 1943 than between 1944 and 1949 must therefore be considered. Two possible causes for the difference are at once apparent. Firstly, a smaller number of children in the earlier age group may have developed cerebral palsy than in the later. Secondly, fewer patients born between 1944 and 1949 who were suffering from cerebral palsy may have died than patients born between 1938 and 1943.

The death rate in cerebral palsy.

The death rate of children with cerebral palsy is unknown. It has been stated that one in seven patients with cerebral palsy dies in infancy. Phelps 1940. Wyllie 1951. Unfortunately, the detailed surveys of patients /

patients on which these estimates are based have not been published, nor have the causes of death been made known. The death rate amongst children with cerebral palsy before adolescence has never been assessed.

In Edinburgh during the past three years three cases of cerebral palsy are known to have died between the ages of 4 and 15. One was a case of bilateral hemiplegia, one of severe diplegia with tetraplegic paresis and one of ataxia. The former two patients died as a result of respiratory infection indirectly due to their cerebral palsy which affected swallowing in both. The third case died as a result of brain stem impaction.

A number of patients in the present series, especially those with severe bilateral palsy, showed impairment of the ability to swallow and in them very numerous respiratory infections occurred. Death as a result of pneumonia would have been inevitable in a number of these patients had antibiotics, especially penicillin, not been available to treat them.

It seems probable therefore that a number of patients with bulbar involvement, chiefly those with severe diplegia or bilateral hemiplegia, born in the antibitotic era are surviving who would have died before this time. This must clearly tend to result in a higher incidence of cerebral palsy amongst children born between 1944 and 1949 than /

than among those born between 1938 and 1943.

A consideration of the possibility of more children born between 1944 and 1949 developing cerebral palsy than those born between 1938 and 1943.

It will be observed from table that the apparently greater incidence of cerebral palsy in the later age group is evident for all forms of cerebral palsy. Without considering aetiological factors in detail it is impossible to discuss the very numerous and complicated factors which may have resulted in a greater proportion of children born in 1944 to 1949 developing cerebral palsy, than children born between 1938 and 1943. Only the more important changes in aetiology which may have resulted in an increased incidence of cerebral palsy will be considered.

Cases of cerebral palsy of congenital aetiology.

It is not possible to give an exact number of cases of cerebral palsy in the present series, in whom the aetiological factors occurred during pregnancy, delivery or the neonatal period. The aetiology of the palsy in some cases is doubtful or unknown. In approximately threequarters of the patients, however, the palsy was of congenital origin. It might be expected that the number of children developing cerebral palsy as a result of prenatal, natal and neonatal causes would alter as maternal and infant care improved. Unfortunately /

Unfortunately, the factors involved in the aetiology of congenital cerebral palsy are so complex that it is almost impossible to determine whether this has occurred. For example, there is a high incidence of birth injury amongst children with hemiplegia. If better antenatal and obstetric care succeeded in reducing the incidence of brain injury fewer cases of hemiplegia might occur. On the other hand because more brain injured infants survive owing to improved medical treatment of the newborn more cases of congenital hemiplegia might also be expected to survive. To what extent improved antenatal and obstetric care is balanced by improved medical treatment in the new born period is difficult to estimate.

Since the infant mortality was actually halved in Edinburgh between 1938 and 1952 it might be expected that some reflection of this great change would be evident in the figures for the incidence of cerebral palsy. Since the reduction of infant mortality is due to so many different causes, however, it is impossible to assess to what degree the incidence has been changed by each. Nevertheless, the reduction in infant mortality does appear to have been accompanied by some rise in the incidence of congenital cerebral palsy. This is particularly evident in cases of diplegia, in which only one of the 79 cases in the present series was thought not to be of congenital origin.

The /

-10-

The greatest single factor in the reduction of infant mortality that occurred between 1939 and 1944 was the reduction in the number of infant deaths that occurred as a result of infections. Crew, 1948, Crosse, 1952. The reduction of the number of infants dying from other causes including prematurity was a less important cause of the total reduction of infant mortality. Nevertheless the neonatal mortality amongst premature infants with birth weights of between 3 and 5½ pounds was reduced by 38.8% approximately in one Edinburgh maternity hospital between 1940-1943 and 1949-1952. Approximately 40% of patients with diplegia in the present series were born prematurely and between 1948 and 1952 the incidence of diplegia amongst prematurely born infants in Edinburgh was approximately 1%. The association of prematurity asphyxia and respiratory infection occurred so frequently in those who later developed diplegia that the association was considered to be of aetiological importance. Clearly, prematurely born infants with respiratory infections have a better chance of surviving infancy than they had before the widespread use of antibiotics and the development of highly developed techniques of infant care.

A relatively high incidence of diplegia must be anticipated in prematurely born babies with respiratory infections /

Table XLIII.

The numbers of diplegic patients and the numbers of prematurely born diplegic patients ascertained in Edinburgh who were born in the years I938-43 and I944-49.

Period of birth	Number of diplegic patients.	Number of diplegic patients born prematurely.
I938-43	22	5
I944-49	45	I9

infections who survive. It is probable that the decreased mortality amongst premature infants since 1943 is an important cause of the increased incidence of diplegia which is apparent since that time. From this point of view it is significant that the proportion of diplegic patients who were prematurely born was twice as great in those born between 1944 and 1949 as it was in those born between 1938 and 1943. Table XLIII.

Acquired cerebral palsy.

The importance of acute infections and their complications in the aetiology of hemiplegia in childhood has long been recognised. Von Heine, 1860, Marie, 1885, Freud, 1897. The importance of septic cerebral thrombophlebitis has only been comparatively recently recognised, however. Symonds, 1937, McAlpine, 1937, Ellis, 1937. Cerebral complications of infectious disease were extremely likely to cause death before the widespread use of antibiotics. Rolleston, 1929. Since sulphonomides and the antibiotics have become more readily available a greater proportion of patients with these complications survive. As a result it might be expected that a higher proportion of cases of acquired hemiplegia would occur in the antibiotic era than before it. The figures in table XLIV. show that there does seem to be a higher proportion of cases of acquired hemiplegia amongst children born between 1944 and 1949 than amongst those born between 1938 and 1943. It seems likely that fewer children with acute /

Table XLIV.

A comparison of the relative proportions of acquired hemiplegia in cases of hemiplegia born between I938-43 and I944-52.

Period.	Number of cases of hemiplegia.	Number of cases of acquired hemi-hemiplegia.	Number of cases of of acquired hemiplegia treated with penicillin.
I938-43	28	8	2[x]
I944-52	47	25	I7
I938-52	75	33	I9

[x] Both developed the infections which were treated with penicillin after I943.

acute infections likely to lead to hemiplegia are dying, and more surviving to develop cerebral damage. That 17 of the 25 patients, suffering from hemiplegia in the later age group, received penicillin, at the time of the acute disease which caused their hemiplegia, indicates the frequency with which pyogenic infections now appear tp cause the condition.

The effects of antibiotic therapy in increasing the incidence of other forms of cerebral palsy are no less startling. Three cases of pyogenic meningitis, one with bilateral hemiplegia, one with ataxia and one with diplegia and cerebellar ataxia would almost certainly have perished without antibiotics. Two cases who had suffered from tuberculous meningitis, one with ataxia and one with diplegia and ataxia, would have died without streptomycin treatment.

Two cases of hydrocephalus who were operated on by modern neurosurgical techniques would not have lived before these were available. One showed ataxia and the other diplegia with ataxia.

If there had been a similar proportion of patients with diplegia born prematurely, a similar proportion of cases of acquired hemiplegia and a similar proportion of patients suffering from the after effects of meningitis, in the two six year periods, the incidence of cerebral palsy amongst those born between 1938 and 1943 /

1943 would have been little lower than for those born between 1944 and 1949.

If, in addition, a number of the patients with cerebral palsy born in the latter period who developed respiratory infections and were treated with antibiotics had died, the incidence of cerebral palsy amongst children born between 1938 and 1943 and between 1944 and 1949 would have been very similar.

It seems probable, therefore, that the incidence of cerebral palsy increased between 1938 and 1949. The increase seems likely to have been due, in part, to a decreased death rate amongst cases suffering from respiratory infections. This appears to have led to a higher incidence of children with severe palsy surviving in the later years. The increase in incidence seems also to have been due in part to a lower mortality occurring in conditions likely to lead to cerebral palsy and especially among premature infants and those with infections in the neonatal period and acute intracranial infections in childhood. The reduced mortality in these conditions has meant that a greater proportion of children with cerebral damage live to manifest cerebral palsy. Much of the apparent increase in the incidence of cerebral palsy seems, therefore, to be attributable to the large scale employment of antibiotic drugs since 1944.

-14-

Comparison of the incidence of cerebral palsy found in the present survey with those found in other surveys.

An attempt has been made to compare the incidence of cerebral palsy obtained as a result of this survey with those found in other surveys of urban areas in England, and with figures obtained from the British Council for the Welfare of Spastics for Wales and England. It will be seen that these vary from 1.0 case of cerebral palsy per 1,000 children of school age to 2.4 cases per 1,000.

The reasons for the variations in the figures for incidence obtained are probably related more closely to the efficiency of ascertainment in the surveys than to differences in the actual incidence of cerebral palsy. But this is by no means as certain as has been suggested by some recent authors. For example, the relationship of the incidence of cerebral palsy of congenital origin to variations in the infant mortality rate is by no means clear. It is likely that places with a relatively high infant mortality rate, and presumably, therefore, a lower standard of maternal and infant care may show differences in the incidence of cerebral palsy from those where the standard of care is higher.

Nevertheless it is probable that the figures in table XLV, for Wallasey, Norwich, Oldham, Salford, Sheffield and Bristol, are roughly comparable to those of the present survey. They were all obtained from urban /

Table XLV.

The incidence of cerebral palsy amongst children of school age in various urban areas of Great Britain.

Authority.	Town.	Incidence of cerebral palsy per 1000.
British Council for the	Wallasey	2.4
Welfare of Spastics.	Norwich	2.0
Selected urban areas.	Salford	2.0
(1948.)	Oldham	1.9
Cohen, H.M.(1953)	Sheffield	1.9
Smallwood, A.L.(1953)	Bristol	2.0
Ingram, T.T.S.(1954)	Edinburgh	2.3

urban areas and were the result of special efforts to ascertain the incidence of cerebral palsy in the populations of school age. It will be seen that they all give an incidence of between 1.9 and 2.4 cases of cerebral palsy per 1,000 children of school age.

Comparison of the incidence obtained in this survey with that obtained in other studies in which fewer sources were contacted.

An attempt was made to make the survey comparable to studies in which only the cases known to public health and school medical authorities and to general practitioners were included. In every case the way in which the patient came to the notice of the survey for the first time was recorded. This is shown in table XLVI. It is thus possible to compare, in a rough way, the incidence obtained in different places where only a proportion of the possible sources of cases of cerebral palsy were contacted.

If one had relied on questionnaires distributed to Edinburgh public health and school medical authorities and to general practitioners, 114 patients with cerebral palsy would have become known. This is equivalent to an incidence of 1.094 cases per 1,000 children born between 1938 and 1952, or approximately half the figure for incidence /

Table XLVI.

The sources of ascertainment in 208 patients with cerebral palsy. The patients with cerebral palsy known to various Edinburgh authorities prior to the survey. The incidence of cerebral palsy in 104,183 children under the age of 15, in Edinburgh when only these cases are considered.

Authority.	Numbers of Patients.	Cumulative Totals.	Incidence per 1000.
School medical service and school for spastics.	97		
Public Health Department.	16		
General Practitioners.	1		
Total known to authorities.		114	1.094.
Hospitals.	52	166	1.59.
Institutions for the mentally defective.	8		
Personal cases.	34		
Grand total.		208	1.991.

incidence found during the more complete survey. The figure of 1.094 patients per 1,000 children in the population agrees very closely however with figures obtained as a result of questionnaires to various unselected urban authorities in England, by the British Council for the Welfare of Spastics. It is similar also to figures obtained as a result of a number of other investigations. TableXLVII Asher and Schonell, 1950. Dunsdon 1952.

Similarly if cases obtained as a result of contacting hospitals are added to those obtained from general practitioners and public health and school medical authorities an incidence for cerebral palsy amongst Edinburgh children of 1.59 cases per 1,000 of the population is obtained. This estimate agrees with that of certain other investigations utilising these sources. Holoran 1952.

On the basis of these comparisons, and more significantly on the comparisons with the more extensive surveys of cerebral palsy in childhood it seems justifiable to state that the overall incidence of the condition appears not to vary very greatly from urban area to urban area in Great Britain.

Comparison of the proportional distribution of cases suffering from various types of cerebral palsy in different surveys.

It /

Table XLVII.

The incidence of cerebral palsy ascertained as a result of Local Authority returns and recent surveys.Schoolchildren.

Authority.	Place.	Incidence per I000
British Council for the Welfare of Spastics.I948.	England.	I.0
Asher and Schonell.I950.	Birmingham.	I.0
Dunsdon.I952.	Various.	I.3
Holoran.I952.	Leeds.	I.6

It is difficult to compare the relative frequency of the different forms of cerebral palsy in different surveys because of the great variations in terminology found in them. In table XLVIII. / an attempt has been made to compare the figures obtained in different investigations. It cannot be regarded as strictly accurate because it has been necessary to depart from the classifications adopted by the investigators themselves and only in some were sufficient details of cases given to make possible reclassification with any confidence. Nevertheless it is probable that the cases in the table give some idea of the frequency of the various types of cerebral palsy in the different surveys.

It will be observed that there are considerable variations in the proportion of cases of each type of cerebral palsy encountered. Thus in the survey of Evans the proportion of dyskinesia is relatively high and the proportion of cases of hemiplegia is low. This is probably because his survey was based on the results of the examination of children who were potential admissions to a special school for cerebral palsy. Since a high proportion of hemiplegic patients can manage at normal schools few might be expected to be seen in his survey. On the other hand the number of patients with dyskinesia who can cope with normal schooling is smaller, and these children, because they tend to suffer less intellectual impairment than /

Table XLVIII.

The proportional distribution of the various types of cerebral palsy, expressed as a percentage, in recent surveys.

	This survey	Asher and Schonell	Evans	Hellebrandt	Dunsdon	
Year	1953	1950	1948	1950	1952	
No. of patients	208	349	112	531	780	
Type of palsy:					Special Schools	Selected areas
Hemiplegia	36.05%	29%	9%	24%	13%	35.3%
Diplegia	37.95%	-	-	-	-	-
Type not stated	-	0.6%	2%	7%	-	-
Paraplegia	13.93%	26%	42%	10%	26%	25.5%
Triplegia	11.06%	5%		4%		
Tetraplegia	12.96%	22%		19%	37%	19.6%
Bilateral hemiplegia	3.86%					
Diplegia and ataxia	5.78%	-	-	-	-	-
Ataxia	7.23%	1%	3%	8%	-	-
Dyskinesia	8.18%	14.4%	40%	27%	13%	7.8%
Other	0.95%	0.4%	-	1%	11%	11.8%
Mixed forms	-	5%	4%	-		
	100%	100%	100%	100%	100%	100%

than those with other forms of cerebral palsy would be encountered. The proportion of cases of dyskinesia was higher and of hemiplegia lower in Dunsdon's survey of children who were candidates for special schools and contrasts with her figures for selected areas, in which cases were unselected.

It is of interest that the figures obtained in this survey are approximately similar to those found in the regional surveys of Asher and Schonell and Dunsdon, in which cases were unselected.

It seems probable that a proportion of cases suffering from the dystonic stage of diplegia were classed as diplegia in the present survey and as dyskinesia in the others. This would clearly tend to result in a higher incidence of diplegia and a lower incidence of dyskinesia in the present survey.

It is interesting that the only survey reporting a comparable proportion of ataxic patients was that of Hellebrandt, which was based on the examination of 531 cases of ataxia in Phelp's clinic. It is possible that ataxia, being the most difficult form of cerebral palsy to diagnose unless the condition is gross, has been missed in a number of patients in the other surveys. Unless ataxia is tested for extremely carefully it is very easy to miss its presence and, as has been noted, a number of ataxic patients /

patients show negative Romberg tests, though they may be quite severely affected. Yet the Romberg test has been regarded as being the soundest and most convenient means of testing for ataxia by many test books on clinical neurology. Monrad-Krohn, 1948.

The incidence of cerebral palsy in the different social classes.

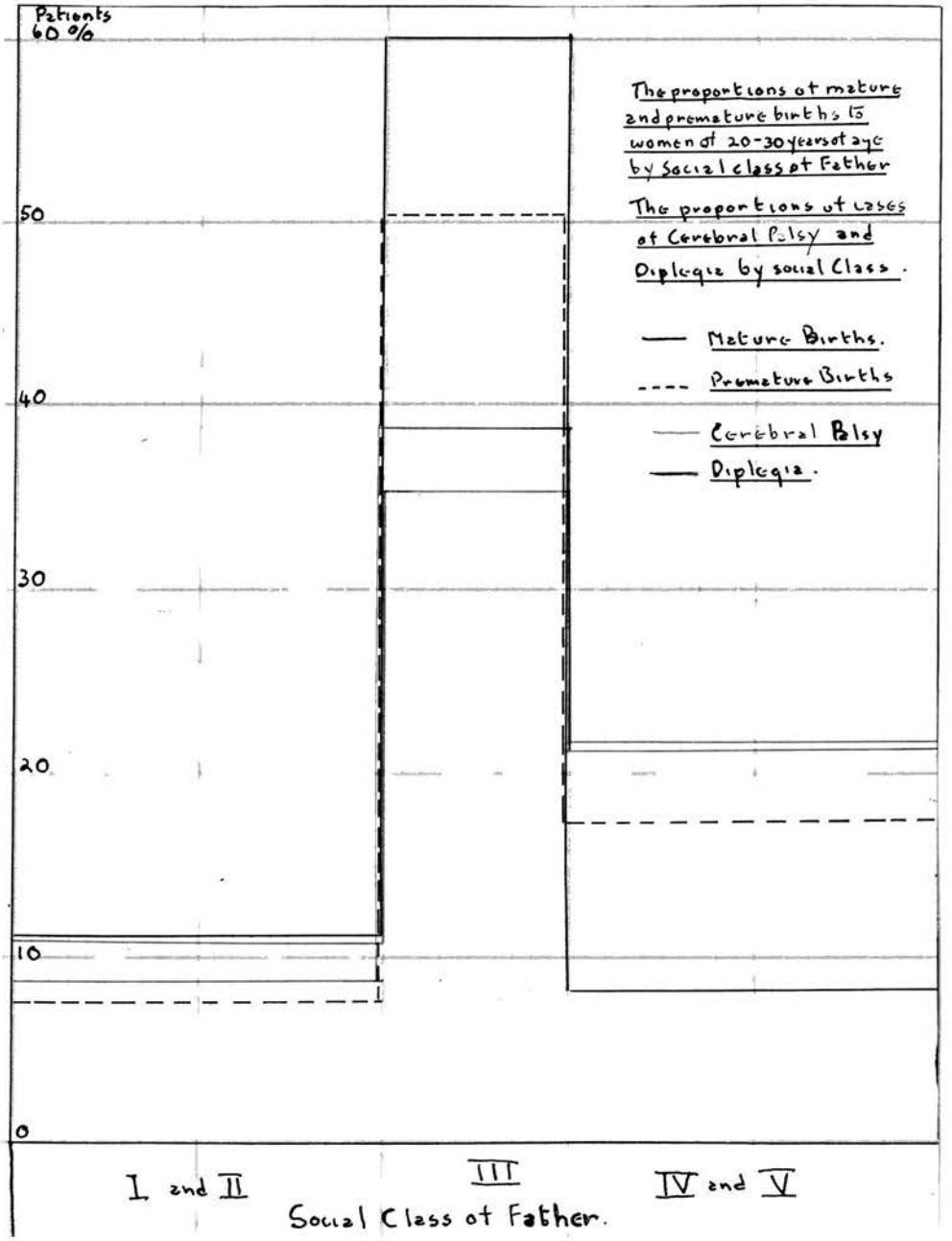
In table IL.is shown the distribution of cases suffering from cerebral palsy of various types. Figures of the distribution of surviving children or of live births by social class of father are based on those obtained by Drillien in a recent sample survey. Drillien, 1954.

It will be observed that there is a comparatively higher incidence of cerebral palsy in the upper and lower social classes than in class III. The reasons for this are multiple, complicated and only partly known. It is apparent that the differences in the proportion of cases in each social class are not the same for all types of cerebral palsy. The proportion of hemiplegic cases in Classes one and two, for example, is similar to that expected on the basis of the distribution of live births. The incidence of patients with diplegia in these classes is higher than expected. All forms of cerebral palsy showed a higher proportion of cases in classes IV and V than expected.

The higher proportion of cases of hemiplegia in classes /

classes IV and V is due largely to an excess of cases of acquired hemiplegia in them. This may be related to the higher incidence of severe infections in childhood, and especially in the early years, in the lower social classes than in the higher. At the same time, it might be expected that if this was the sole factor operating, the incidence would be lower in classes one and two. That this is not so may be an accidental finding, or it may be due to the better medical treatment of severe infections in childhood in them, resulting in a higher proportion of brain damaged children surviving.

There is a higher proportion of cases of diplegia in classes I and II and IV and V than in class III. Since a high proportion of cases of diplegia are first born or only children a higher incidence of the condition might be expected in classes I and II than in classes IV and V in which the families tend to be larger. The maternal ages in cases of diplegia were known and from them and a consideration of the maternal age in all live births in the various social classes it was possible to apply correcting factors to the expected incidence of diplegia in each. The correcting factor was very small; 2% to be added to the number of expected cases in classes I and II and 2% subtracted from the number of expected cases in class V. The effect of the higher maternal age in classes I and II compared /


Patients
60 %
50
40
30
20
10
0
The proportions of mature and premature births to women of 20-30 years of age by Social class of Father
The proportions of cases of Cerebral Palsy and Diplegia by social Class.
Mature Births.
Premature Births
Cerebral Palsy
Diplegia.
I and II
III
IV and V
Social Class of Father.

compared to the other social groups was therefore a factor of small importance in raising the incidence of diplegia in them.

The higher proportion of cases of diplegia in social classes IV and V than in classes I, II and III appeared to be largely due to a higher proportion of prematurely born patients with diplegia in the former. It was possible to obtain figures for the proportion of premature births by social class of father from a recent Edinburgh survey. Table L. Drillien, 1954. It will be seen that the difference in the proportion of cases of diplegia in classes IV and V and in the others is greater for diplegia than for premature birth however. It seems unlikely, therefore, that the greater proportion of premature births in classes IV and V is the only cause of the higher incidence of diplegia in these classes. A greater incidence of neonatal respiratory infections in classes IV and V and poorer care of the premature infant are other possible contributory causes of the higher incidence of diplegic paresis.

No figures are available on which to base an assessment of the effects of differences in the incidence of abnormalities of pregnancy and delivery in the social classes on the proportion of cases of diplegia which occurred in each.

The /

Table L.

The numbers of patients with cerebral palsy and their proportions by social class. The social class incidence of a group of premature infants compared to a group of mothers giving birth to mature infants. All mothers between 20 and 30 years. Still births and infant deaths excluded. Drillien 1954.

Class of Father.	Mothers between 20 and 30 years of age. Mature group. %	Premature group. %	Cerebral palsy. Mothers of all ages, not only from 20-30. Diplegia. Number.	%	Hemiplegia. Number.	%	Other forms. Number.	%	All forms. Number	%
I	22.7.	14.5	5	6.33	2	2.66	4	7.5	11	5
II			12	15.19	6	8.0	7	12.9	25	1
III	60.2	50.9	28	35.44	29	38.66.	24	44.4	81	3
IV	17.1.	34.6.	17	21.52	18	24.0	8	14.8	43	20
V			17	21.52	20	26.66	11	20.4	48	22
All classes.	100	100	79	100	75	100	54	100	208	10

ality into good, intermediate and poor wards.
infant mortality and infectious diseases from Edinburgh Public Health reports.

Cerebral palsy.	Incidence per 1000 under 16. Hemiplegia.	Diplegia.	Cerebral Palsy.	All ages. Incidence of infectious disease totals for 1950,51,52. Per 1000 Scarlet Fever.	Whooping cough.
15	1.221	.814	3.053	17.301	64.540
12	.652	.652	1.955	26.389	106.206
14	.716	1.194	3.343	19.819	46.323
17	1.067	.534	1.814	16.539	50.256
9	1.215	.912	2.735	17.320	50.137
8	.888	.178	1.421	23.091	31.972
14	.898	.898	2.515.	25.153	28.566
6	.268	.804	1.609	11.796	50.134
12	.651	1.085	2.603	24.078	55.748
107	.860	.735	2.255	20.22	54.33
11	1.593	.797	2.923	22.582	58.198
6	0	.606	1.817	27.248	56.010
3	.301	.602	.902	25.526	50.526
7	.531	.797	1.860	22.317	49.416
12	1.309	.748	2.244	20.572	54.984
5	.234	.701	1.169	14.964	51.438
14	.789	1.381	2.761	21.696	47.140
4	0	.770	1.539	15.006	30.012
60	.672	.832	1.909	21.33	50.85
7	1.767	.707	2.474	25.097	37.116
5	0	1.158	1.931	23.938	28.958
14	.549	.961	1.922	14.280	31.855
7	.452	.452	1.583	32.783	26.905
6	.604	.604	1.813	18.132	31.732
2	0	.868	.868	18.229	40.799
41	.52	.72	1.803	19.35	27.99
208	0.735	0.704.	2.046		

Table LI.

The incidence of cerebral palsy in Edinburgh wards grouped according to the in
Figures for population obtained from census returns of the Registrar General.F

	Ward.	Infant mortality. Average I950,5I,52.	Population. aged 0-I5 years.	Number of Hemiplegia	Di
Poor wards. Infant Mort. 3I plus per I000.	St,Giles.	34	4,9I3	6	
	Liberton.	35	6,I39	4	
	Colinton.	3I	4,I88	3	
	Pilton.	33	9,372	I0	
	St.Bernards.	3I	3,29I	4	
	Portobello.	3I	5,630	5	
	Craigmillar.	40	5,566	5	
	Calton.	32	3,730	I	
	Holyrood.	32	4,6I0	3	
Totals and averages for poor wards. Average population per ward under I6 years of age.5,27I.		33.2	47,439	4I	
Intermediate Wards.Infant mortality between 26 and 3o/I000	Newington.	29	3,763	6	
	St.Andrews.	29	3,3o3	0	
	Broughton.	26	3,325	I	
	West Leith.	27	3,764	2	
	Central Leith.	25	5,347	7	
	South Leith.	27	4,277	I	
	Craigentinny.	27	5,070	4	
	Merchiston.	30	2,599	0	
Totals and averages for interemdiate wards.Average population under I6. 302I.		27.5	3I,448	2I	
Good wards. Infant mort. under 25.	George Square.	2I	2,829	5	
	Morningside.	I4	2,590	0	
	Sighthill.	I7	7,283	4	
	Gorgie/Dalry.	I7	4,423	2	
	Corstorphine.	2I	3,309	2	
	Murrayfield/Cramond.	23	2,304	0	
Totals and averages for good wards. Average population under I6.3790.		I9.9	22,738	I3	
City of Edinburgh.Figures based on I95I census returns.		28.3	I0I,625	75	

The higher incidence of other forms of cerebral palsy than expected in classes I and II is largely due to an excess of patients suffering from dyskinesia. It is difficult to say why there should be a higher proportion of these patients in the upper social classes than in the lower. Possibly the care of infants suffering from the effects of rhesus incompatability was better in classes I and II and a higher proportion of brain damaged children survived.

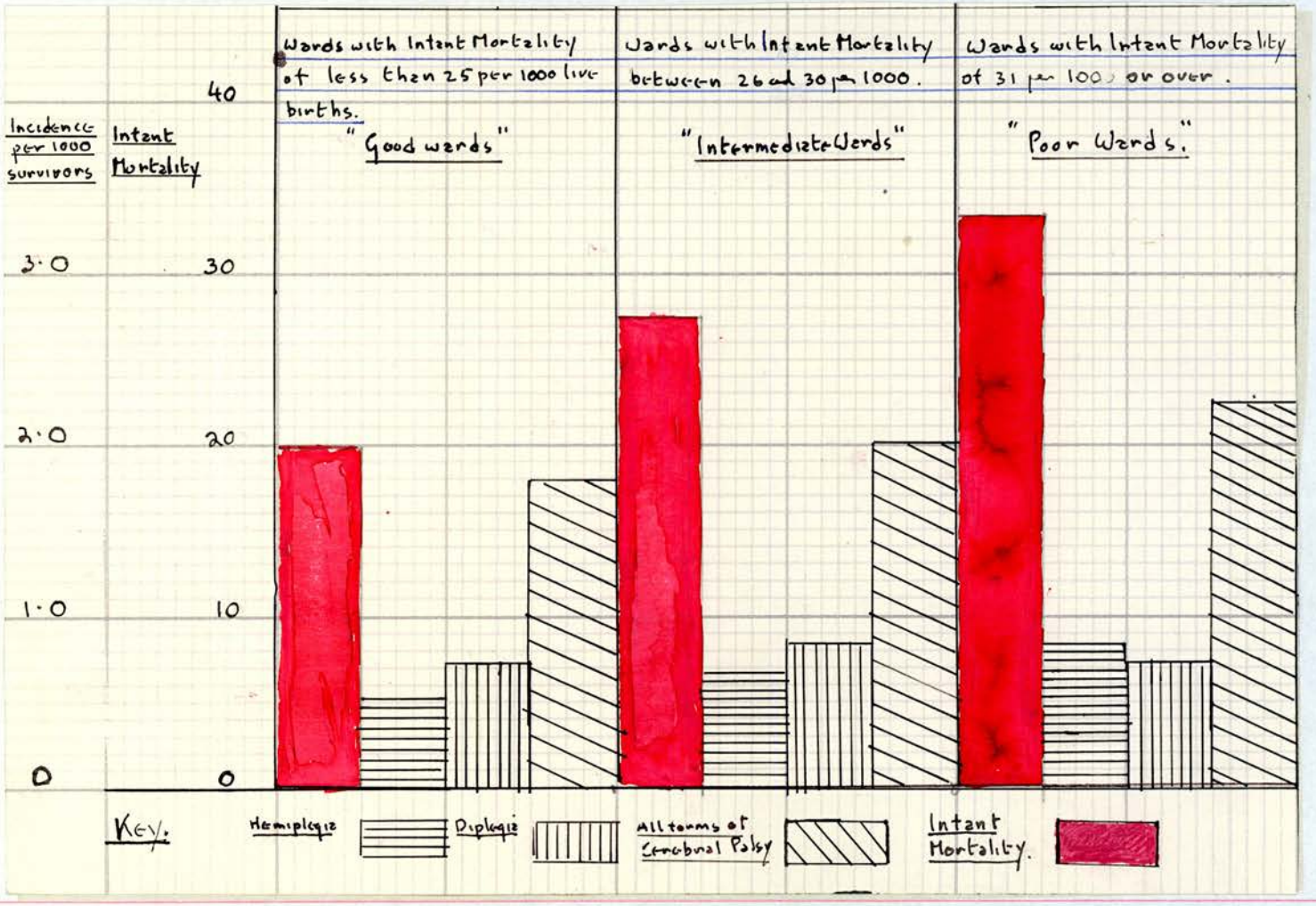
The incidence of cerebral palsy in Edinburgh wards.

An attempt was made to assess the incidence of cerebral palsy in childhood in different districts of Edinburgh. This was done for two reasons; firstly, to confirm that ascertainment was relatively uniform throughout the city and, secondly, to investigate possible variations of incidence between better class and poorer districts.

The decision as to whether a district was a better class one or a poor one was arbitarily based upon its infant mortality for the years 1950, 1951 and 1953. Wards with an infant mortality of under 25 were classified as better class wards, those with an infant mortality of 31 or over as poorer class wards and those with an infant mortality of between 26 and 30 as being intermediate. In table LI. are shown the figures for the incidence of cerebral palsy in each ward together with figures of the

numbers /

TABLE LII.



The incidence of cerebral palsy compared to the infant mortality in Edinburgh wards.

numbers of people per room, incidence of measles, scarlet fever and whooping cough and infant mortality.

It will be seen that the incidence of cerebral palsy varied from .868 to 3.343 cases per 1,000 children aged 15 and under in the different wards but that cases of cerebral palsy were found in all. It seems unlikely that the survey was grossly uneven in the efficiency of its ascertainment in different wards. In general the wards with a low incidence of cerebral palsy were those classified as better class wards, those with a high incidence had been classified as being poorer class wards.

That living conditions do appear to have some effect on the incidence of cerebral palsy is evident from a study of table LII. It will be seen that the incidence of cerebral palsy is significantly higher in poor wards than in intermediate wards and in the latter than in better class wards. The incidence of all forms of cerebral palsy does not conform to this pattern, however, as diplegia appears to occur more commonly in the intermediate group than in the better class wards or the poorer wards. The reasons for this are obscure. It is possible that the main factors determining the incidence of diplegia are the incidence of premature birth, higher in social classes IV and V and the proportion of only children occurring in the /

the community, higher in classes I and II. In the good wards the effect of the higher proportion of children may be counterbalanced by the lower incidence of prematurity. In the poorer wards the effect of the higher incidence of prematurity may be balanced to some extent by the smaller number of only children. In intermediate wards, however, both these factors may be important causes in different social classes in them, and the balancing effect of each factor upon the other may be lost. It is interesting in this connection that the average child population in both the good and the intermediate wards is between 3,500 and 4,000, whereas the average population in the poor wards was over 5,000. The variations in adult populations in the various wards were much less marked. The birth rate in the better and intermediate wards was smaller and the proportion of only children was greater than in the poorer wards. The relatively small effect of maternal age on the incidence of diplegia in the different social classes has been referred to and seems unlikely to have affected the incidence of the condition in the various wards to any great extent.

The incidence of congenital and acquired hemiplegia was significantly higher in the intermediate wards than in the good wards and in the poor wards than in the intermediate. This may be related to the greater incidence of acute /

acute infections during childhood in the poorer wards causing a greater proportion of cases of acquired hemiplegia. The somewhat greater incidence of congenital hemiplegia in the poorer and intermediate wards than in the good wards may be related to a lower standard of medical care during pregnancy and delivery. Without considering aetiological factors in detail it is impossible to assess to what degree variations in the standards of maternal care have affected the incidence of congenital hemiplegia in the different wards.

The number of cases in each of the other categories of cerebral palsy was relatively small. Variations in incidence were very likely to be the result of chance and no statistical consideration is possible. When all the other forms of cerebral palsy are considered together, the incidence is found to be significantly increased in poor wards compared to intermediate and in intermediate compared to good wards.

The difficulties in drawing conclusions about the causes of the differing incidence in the various types of cerebral palsy in the different wards is obvious. The aetiological causes are multifactor and interrelated. To assume that any one cause is responsible for differences in the incidence of any single type of cerebral palsy is extremely liable to be fallacious, unless the effects of numerous /

numerous other factors are borne in mind. Nevertheless it seems certain that the incidence of cerebral palsy tends to be higher where social conditions are poor, and lower where social conditions are good.

CHAPTER 2

The significance of cerebral palsy in childhood to the community.

In this chapter an attempt is made to describe the incidence of the major disabilities suffered by the children with cerebral palsy discovered during the survey. Bare figures giving the incidence of the various forms of cerebral palsy give only a very imperfect impression of the severity of the handicaps the condition causes. What follows is designed to elaborate the incidence figures given in the previous chapter. The incidences of the more frequent and severe handicaps suffered by children with cerebral palsy are discussed and a brief consideration is given to the effects they have on the child's education and prospects of employment.

The sex distribution of children with cerebral palsy.

In the series of 208 children with cerebral palsy, 126, or 60.5% were males and 82, or 39.5% were females. This distribution is similar to that reported by a number of recent surveys. Asher and Schonell, 1950. British Council for the Welfare of Spastics, 1948. In other surveys a more equal distribution of cerebral palsy between the sexes is reported. Cohen, 1953.

Females outnumbered males in only two forms of cerebral palsy, bilateral hemiplegia and dyskinesia. The male preponderance means that a higher proportion of potential /

potential wage earners are affected than if the distribution of cerebral palsy had been more equal. On the other hand, the condition is possibly a greater handicap to the female who is affected. Her chances of marrying are small and her chances of obtaining remunerative employment in the more highly competitive female labour market are probably less than those of a similarly affected male. Table LIII.

The severity of the palsy.

Because of the diversity of the manifestations of the various types of cerebral palsy it was necessary to use different criteria to assess the severity of the disorder in each type. In some ways this was unfortunate for it is apparent that a child with severe hemiplegia for example in which one hand is functionally normal is less severely affected from the points of view of education and employment than a child with tetraplegia who may be only moderately severely affected. It is possible to take account of the extent of the disability as well as of its severity, however, and thus to obtain some idea of the total physical handicap suffered by the child. In general it is true of all types of palsy that patients classified as being slightly affected are able to take part in normal activities, those with moderately severe palsy will have their activities somewhat curtailed and

Table LIII.

The distribution of the various forms of cerebral palsy by sex.

	Hemiplegia.	Diplegia.	Ataxic Diplegia.	Ataxia.	Dyskinesia.	Bilateral Hemiplegia.	Other.	Total.	Approximate. Percentage.
Male.	52	49	7	9	7	I	I	I26	60.5
Female.	23	30	5	6	I0	7	I	82	39.5
Totals.	75	79	I2	I5	I7	8	2	208	I00

patients with severe palsy will be largely incapacitated. Table LIV.

In the present series of 208 patients, 52 or 25% were classified as being slightly affected. In them there was some impairment of function of the affected limbs but normal activities were not limited to any great extent by the palsy. In many of these patients the most severe manifestations of their palsy were evident in the early years. Motor development was retarded. Because the child walked later, used his hands later and could wash and dress himself later than the normal child more and closer attention had to be paid to him in his prolonged infancy than to a normal child. Unless other handicaps were present most mildly affected children were able to benefit from normal education and could be employed in a variety of jobs with normal people later. Normal activities might be clumsy and rather slow but most were possible and the child could adjust relatively successfully to his disability if his intelligence was unimpaired.

80 children, or 38%, were classified as being moderately severely affected by cerebral palsy. In them more prolonged care was necessary in the early years than in mildly affected children. The age at which moderately affected patients walked, dressed, fed and washed themselves was later than in mildly affected patients. Some require /

Table LIV.

The severity and extent of cerebral palsy in 208 patients.

The severity of the palsy.

Type of Palsy.	Number of mild cases.	Moderately severe.	Severe.	Totals.
Hemiplegia.	27	26	22	75
Diplegia.	I3	35	3I	79
Diplegia and ataxia.	4	4	4	I2
Ataxia.	4	8	3	I5
Dyskinesia.	4	6	7	I7
Bilateral hemiplegia.	0	0	8	8
Other.	0	0	I	2
Totals.	52	80	76	208
Percentage distribution.	25	38	37	I00

The extent of the palsy.

Type of palsy.	Two useful upper limbs.	One useful upper limb.	No useful upper limb.	Totals
Hemiplegia	27	48	0	75
Diplegia	33	I9	27	27
Diplegia with ataxia	4	6	2	I2
Ataxia	I5	0	0	I5
Dyskinesia	3	3	II	I7
Bilateral hemiplegia	0	0	8	8
Other	I	0	I	2
Totals.	83	76	49	208
Percentage distribution.	40%	37%	23%	I00

require help with ordinary activities, such as dressing until the age of 9 or 10. Walking was or will be possible in all the moderately severely affected patients but in some its development is very late and the gait is unsteady. There was at least one functionally useful limb in all the moderately severely affected patients but in 9 there was some bilateral impairment of fine manipulation. In 45 moderately severely affected patients one upper limb showed functional impairment sufficiently severe to render many normal activities, especially feeding, impossible with the affected arm. In 26 patients both upper limbs were spared or affected insignificantly, so that most normal activities were possible with both arms. The degree to which the arms were affected was frequently of crucial importance in affecting the child's ability to benefit from education and later to take employment. Though it was possible to wheel a child to school if his walking was retarded, unless he had one useful upper limb, it was a matter of extreme difficulty to teach him to write or learn to manipulate. Some of the patients with moderately severe cerebral palsy may be able to work in jobs which are adapted to their disability, though few are capable of manual work. In present circumstances, without special provision being made for them, the majority would be unemployed.

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Severe cases of cerebral palsy numbered 76 or 37% of the series. In them a degree of care is necessary for a very long period of childhood and in the majority supervision and help will be necessary for life. Approximately one third of the patients cannot and are not likely to be able to walk. In the patients who will be able to walk the gait will be unsteady and grossly abnormal in all except those with hemiplegia. The hemiplegic patients were classified as severe because their affected upper limb was almost completely useless functionally in all cases.

Five of the 76 patients considered to be severely affected were able to use both upper limbs to some extent though there was neurological involvement in all of them. They were classified as being severely affected because in all walking was grossly defective. In 31 patients one limb was functionally useful and in 40 normal manipulative ability was impossible with either hand.

In all the patients with severe palsy special educational facilities are required and the majority require special care in addition, in order that they may develop at least some independence in dressing, washing, and feeding themselves. In present conditions only very occasional cases of severe cerebral palsy could be found suitable jobs without special facilities for their care being provided. The majority are unemployable.

Speech /

The number of patients found to be suffering from a number of major disabilities among 208 Edinburgh children with cerebral palsy.

Table LV.

Classification of Palsy.	Upper Limbs. Two Useful	One Useful	None Useful	Intelligence IQ.70& under85	I.Q. under70	Epilepsy. Grand Mal Jacksonian	Petit Mal other	Aphasia - Aphasia	Visual Defect	Speech Defect	Over Activity	Schooling. Normal School	P.H. School	M.H. School.	Ineducable	Not yet at School	No. of cases
Hemiplegia																	
Mild	27	0	0	4	11	6	0	2	1	11	3	9	2	8	4	6	27
Mod. Severe	0	26	0	4	11	10	4	6	4	8	4	6	1	5	1	7	26
Severe.	0	22	0	9	4	8	0	4	6	2	2	8	5	2	4	4	22
TOTAL	27	48	0	17	26	22	4	12	11	21	9	23	11	15	9	17	75
Diplegia.																	
Paraplegia	29	0	0	6	8	6	1	0	2	7	0	7	9	5	0	8	29
Triplegia.	4	19	0	5	6	9	3	3	3	14	0	3	3	5	4	8	23
Tetraplegia	0	0	27	4	14	7	5	1	8	14	0	1	0	6	9	11	27
TOTAL.	33	19	27	15	28	22	9	4	13	35	0	11	12	16	13	27	79
Diplegia with ataxia																	
Triplegia	3	6	0	0	1	1	2	0	0	8	1	2	3	1	2	1	9
Tetraplegia	1	0	2	1	3	0	2	0	1	2	0	0	1	1	0	1	3
TOTAL	4	6	2	1	4	1	4	0	1	10	1	2	4	2	2	2	12
ATAXIA.	15	0	0	6	3	3	1	0	2	11	4	2	7	3	0	3	15
DYSKINESIA	3	3	11	4	2	0	4	0	0	15	1	2	7	2	1	5	17
Bilateral Hemiplegia	0	0	8	0	8	5	0	0	8	8	1	0	0	0	8	0	8
Other forms of cerebral palsy	1	0	1	1	0	0	0	0	0	2	0	0	2	0	0	0	2
Grand Total	83	76	49	44	71	53	22	16	35	102	16	40	43	38	33	54	208
	39.8	36.5	24.5	21.1	34.1	28.4	10.5	7.7	16.8	48.8	7.7	19.3	20.7	18.2	15.9	25.9	

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Speech defects in children with cerebral palsy.

The crude criterion used for deciding whether a child had defective speech in the present series was whether comprehension of his speech was rendered difficult by reason of its being abnormal, or not. Minor speech defects which were not sufficient to impair comprehension of the child's speech were ignored. Generous allowance was made for some retardation of speech in those under the age of 4 because a high proportion were mentally defective, and in them some degree of retardation and infantile speech was almost always present.

102 patients or 48.8% of the present series were considered to suffer from speech defects severe enough to result in difficulty in understanding what they said. This compares with the figure obtained in one recent survey, of 79% for all speech defects in children with cerebral palsy of school age. Dunsdon 1952. Since this figure includes cases with those minor defects which were ignored in the present survey, it can probably be regarded as comparable.

In table LV. are shown the numbers of patients suffering from each type of cerebral palsy who suffered from significant speech defects. It will be observed that dyslalia of various types accounted for the main difficulty in speech in 30.8% of patients with cerebral palsy. 10.5% of patients had no more than one or two words of comprehensible /

comprehensible speech. This figure may be compared to the figure for loss of speech of 16% quoted by Dunsdon. Her 16% appears to have included a number of cases of aphasia, however, which numbered 7.7% of the present series. Taking account of this it is probable that the figures for very severe loss of speech are comparable.

Speech defects were more frequent in patients with severe cerebral palsy than in those with mild cerebral palsy and in those with mental defect than in those with normal intelligence. The incidence was least in patients with hemiplegia and greatest in patients with bilateral hemiplegia, dyskinesia and ataxia.

In this series 24.5%, including the 10.5% with not more than one or two words, were considered to show severe speech defects so that they were incapable of expressing ideas in a fashion likely to be understood without patience on the part of both the children and their hearers. The majority of these patients were severely handicapped physically and were mentally defective. Less than one quarter were of approximately normal intelligence, approximately 5% of the whole series of patients with cerebral palsy. It is in this 5% of patients that speech defects cause important handicaps, impair school performance and later reduce the chances of the child finding employment. In a number of these children with severe

speech /

speech defects and relatively unimpaired intelligence the speech defect was a handicap of sufficient severity to determine their admission to physically handicapped instead of to normal schools.

Hearing defects.

Deafness of a degree severe enough to necessitate education as a deaf child was present in only one case. Seven patients with dyskinesia were shown to suffer from high frequency deafness as a result of audiometry and another two were suspected of having high frequency deafness but were too young for testing.

Four patients in the survey were hard of hearing, one as a result of meningitis and the other three as a result of chronic middle ear disease.

Visual defects.

Visual defects not due to refractive errors were encountered in 35 patients, in the present series. The incidence of visual defects was much higher in patients with severe cerebral palsy than in those with mild cerebral palsy. They were frequently associated with marked mental defect, as in all the eight patients with bilateral hemiplegia. They were therefore rather less important as a severe cause of restriction of activities than would otherwise have been the case. In seven patients considered to be /

be of educable standard the visual defects are sufficiently severe to necessitate education by non visual means now or in the future. Four of these patients were cases of diplegia with retrolental fibroplasia under the age of 5.

The very good compensation achieved by some patients with field defects was remarkable. It was interesting to observe that two patients with hemianopia who had both had a number of medical examinations had not been noted to have any visual defect until this was demonstrated during the survey.

Epilepsy.

In table LV.are shown the numbers of patients who suffered from epilepsy of various types. 16 patients suffered from petit mal or similar attacks involving brief impairment of consciousness. Some of these patients also suffered from grand mal. Petit mal occurred more frequently in those with severe or extensive palsy than in those with mild disability. It was therefore a nuisance to the patient rather than a serious cause of limitation of activities for most of those affected were unemployable in any case.

Grand mal and attacks of Jacksonian type with loss of consciousness occurred in 53 patients, or approximately 25%. The frequency of the attacks varied greatly as was noted /

noted when the various forms of palsy were discussed. In the majority, however, the attacks occurred between once a month and once a year in spite of antiepileptic drugs. The proportion of moderately severely and severely affected patients with grand mal epilepsy was only slightly higher than of those with mild cerebral palsy. There was a tendency for attacks to become less frequent with age. In view of this it is difficult to give any assessment of the frequency with which the choice of jobs for the affected child would be further limited because he was epileptic. Epilepsy was the deciding factor in sending mildly affected children to schools for the physically handicapped rather than to normal schools in a number of patients.

Overactive behaviour.

Sixteen patients with cerebral palsy showed over-activity which was characterised by an intense preoccupation with the immediate surroundings, gross impairment of concentration and of attention span and almost complete lack of common sense or insight. The patients were never still. All objects in a room, furniture and people attracted the child's intense superficial interest momentarily. Objects were handled, frequently put to the mouth and chewed and then discarded. Interest in anyone was only sustained for a matter of a few seconds or a couple of minutes at the most. Nine of these patients were cases of hemiplegia and 4 were ataxia. The behaviour was a tremendous handi-

cap to the child's social development and education, as owing to the grossly defective attention span and lack of insight nothing, or very little, could be taught.

The overactivity appeared to be most marked up to the age of 8 or 10 and then to decrease in severity, the child gaining some insight, and a gradual increase in the length of his attention span becoming apparent at the same time. Only four of the patients were considered educable, however, and the progress of all had been poor.

The overactivity is similar in type to that described in epileptic patients by Peterman 1953. In severe cases it bore a striking resemblance to the overactive behaviour disorder found in monkeys as a result of temporal lobe lesions by Kluver and Bucy, 1939.

Intellectual impairment in cerebral palsy.

The majority of patients with cerebral palsy are of subnormal intelligence. This has been the conclusion reached as a result of a large number of recent surveys. It is borne out to some extent in the present series. Since the intelligence quotients shown are the result of a number of different tests, performed by a variety of testers in unknown circumstances, it is unwise to regard the results as very accurate. Because the degree of their accuracy is unknown it has been felt desirable to quote a number of more systematic surveys of the intelligence of children /

Table LVI.

The intelligence quotients of 208 patients with cerebral palsy. Percentage distribution.

Intelligence quotient. / Type of palsy.	Over 115	100-114	85-99	70-84	55-69	Under 55	Untested.	Totals.
Hemiplegia.								
Mild.	I	2	4	4	8	5	3	
Mod.severe.	2	4	2	4	7	4	3	
Severe.	0	4	4	9	2	2	I	
Total.Hemiplegia .	3	I0	I0	I7	I7	II	7	75.
Diplegia.								
Paraplegic.	I	5	4	6	7	I	5	
Triplegic.	2	2	0	5	3	3	8	
Tetraplegic.	0	0	2	4	3	II	7	
Total diplegia.	3	7	6	I5	I3	I5	20	79.
Diplegia with ataxia.	0	2	3	I	I	2	3	I2.
Ataxia.	0	I	3	6	3	0	2	I5.
Dyskinesia.	I	2	2	4	2	I	5	I7.
Bilateral hemiplegia.	0	0	0	o	0	8	0	8.
Other.	I	0	0	0	I	0	0	2.
Totals.	8	22	24	43	37	37	37	208.
Percentage distribution.	3.8	I0.6	II.5	20.7	I7.8	I7.8	I7.8.	I00.
Percentage distribution of those tested.	4.9	I2.8	I4.3	24.2	2I.9	2I.9.	-	I00.

children with cerebral palsy. TableLVI. The results of the other tests are not dissimilar, however, to those obtained during the present survey.

It will be observed that there is a higher proportion of patients with normal intelligence in the present series than in most others. This is probably the result of more adequate ascertainment of relatively mildly affected children in normal schools than in other surveys. There is also a higher proportion of grossly defective children in the present series. This is probably the result of fuller ascertainment of cases of cerebral palsy in institutions for the mentally defective. Table LVII.

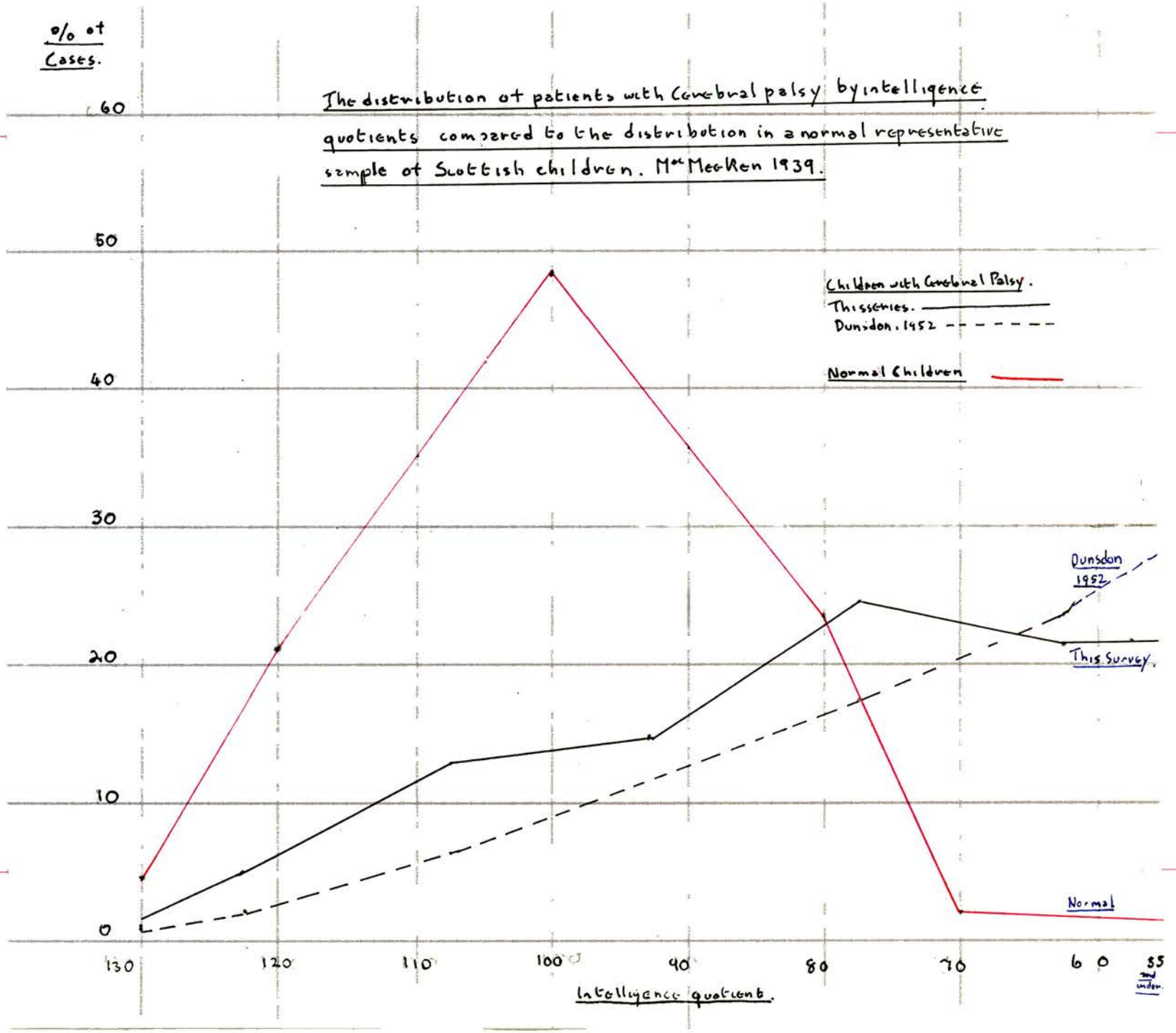
Thirty seven patients in the present survey had not been tested psychologically. Twenty one of these were under the age of three years. The remaining 16 patients were not tested for a variety of reasons but there seemed to be no predominance of patients in the ineducable, normal school or special school groups in these. The 171 patients who were tested may probably be regarded as being representative of the present series. 43.8% of these children had intelligence quotients of 69 or less. They were therefore in need of special educational provision, whatever the severity of their cerebral palsy. The educational progress of most of these patients is likely to be small. For practical purposes the combination of mental defect and cerebral palsy of whatever severity renders /

Table LVII.

A comparison of the percentage distribution of intelligence quotients in this survey with that found in other recent investigations, and in a group of unaffected Scottish children

Intelligence quotients	130 or more	115 to 129	100 to 114	85 to 99	70 to 84	55 to 69	Under 55	
Dunsdon 1952	0.5	2	6.2	15.5	17.2	23.6	35	
This survey	4.9		12.8	14.3	24.2	21.9	21.9	

Intelligence quotients	130 or more	110 to 129	90 to 109	70 to 89	50 to 69	25 to 49	Under 25	Untest-:able
Asher and Schonell 1950	0.6	3.4	20.1	26.8	22.9	10.7	11.6	3.9
Holaran 1952	-	2.8	22.0	36.2	21.3	8.5	3.5	5.7
Unaffected children MacMeeken 1939	4.7	21.2	48.6	24.3		1.3		


% of Cases.
The distribution of patients with Cerebral palsy by intelligence quotients compared to the distribution in a normal representative sample of Scottish children. Mac Meeken 1939.
60
50
40
30
20
10
0
Children with Cerebral Palsy.
This series.
Dunsdon. 1952
Normal Children
Dunsdon 1952
This Survey.
Normal
130
120
110
100
90
80
70
60
55 and under
Intelligence quotient.

renders them completely unemployable except under institutional supervision.

43 children, or 24.2% of the tested patients, had intelligence quotients of between 70 and 84. A few of these patients with mild cerebral palsy were attending normal schools but the majority were in schools for the mentally or physically handicapped and were in need of special educational methods. It is probable that a few of the mildly affected patients are employable but the prospects for remunerative employment are very small for the moderately severely and severely handicapped children.

54 children, 32% of those tested, had intelligence quotients of 85 or more and may, charitably in some cases, be regarded as of normal intelligence. Approximately two thirds of these patients were at normal schools but the remainder were in schools for the physically handicapped. Those in the latter included the majority of those with severe physical disability.

Aphasia.

Aphasia was evident in 16 cases of cerebral palsy, or 7.7% of the patients in the series. 12 of the patients were hemiplegic and eleven showed right hemiplegia. All but two of the 16 cases were moderately severe or severe cases of cerebral palsy and some degree of mental defect was present in 9. In all, however, the degree of verbal

difficulty /

difficulty was out of all proportion to the degree of mental defect. Cases with severe mental defect in which there seemed to be some degree of specific difficulty in expression were regarded as cases of mental defect and not of aphasia. It is probable, therefore, that full psychological testing would have resulted in a higher proportion ofaphasic patients than was noted during the survey.

The most marked effects of aphasic disorder were apparent in the early years at school where great difficulty in learning to read and write were encountered and were not recognised as being due to specific organic disability in most cases. Since the presence of aphasia results in a much lower score in verbal intelligence tests than would otherwise be the case, the intelligence quotients for aphasic children must be accepted with the greatest reserve. Most of those in the present series had been tested by routine intelligence tests by testers unaware of the significance of aphasia and these results were downright misleading. Retesting was necessary in all aphasic patients in the present survey.

Specific learning difficulties in children with cerebral palsy.

In addition to the handicaps imposed on the child's ability /

ability to learn as a result of motor disabilities, mental impairment, visual, hearing and aphasic defects, certain other specific educational difficulties occurred frequently in patients with cerebral palsy. Table LVIII.

A number of patients appeared to have the greatest difficulty in recognising letter and word shapes. In children in whom the difficulty was not marked it was most frequently shown in a tendency to reverse small words and to confuse letters with their mirror image. Thus "dog" would be read and written instead of "god", "was" instead of "saw". The printed letters "b" and "d" and "p" and "q" were frequently confused. In patients with more severe difficulty of this type whole lines would be read in a mechanical fashion, or written in reverse. The difficulty was much less marked with larger words, probably because of their more distinct shapes. Some children would read quite complicated passages guessing the small words or ignoring them and managing the large words quite well.

When the difficulty was mild it frequently affected only writing, but when it was severe it affected both reading and writing. In most of the affected patients with hemiplegia or diplegia, reversal of words and letters occurred in only one dimension, most commonly the horizontal. In patients with severe ataxia on the other hand, bizarre plays in both the vertical and horizontal /

Table LVIII.

The number of patients with cerebral palsy between the ages of 6 and II who were found to reverse letters when reading or writing in a small Edinburgh survey.

Type of palsy	Number of patients.	Reading only.	Writing only.	Both reading and writing.	Totals
Right hemiplegia.	I2	I	4	3	8
Left hemiplegia.	I3	0	I	I	2
Diplegia.	24	0	3	2	5
Ataxia and diplegia with ataxia.	5	0	2	0	2
Dyskinesia.	3	0	0	0	0
Totals.	57	I	I0	6	I7.

zontal dimensions were evident.

In most patients with cerebral palsy the reading and writing difficulties were evident from the age of 6 to 8 and persisted for a variable time. One boy of average intelligence still reverses small words in reading and writing at the age of 14. In most cases the reversals are only evident for a matter of months, however.

Reversals of letters and small words occur fairly commonly in normal children in primary school as transient phenomena lasting for a few weeks and for longer in mentally defective children. Orton, 1937. Many attempts have been made by Orton and others to relate lateralising difficulties to mixed cerebral dominance and latent sinistrality in the apparently right handed. MacMeeken, 1939. The difficulties have also been related to defects of the ability to conceive spatial relationships shown by some of the children affected. It has been suggested that the reason that lateralising difficulties are more frequent and severe in mentally defectives is that their spatial concepts are less well formed than in normal children.

Lateralising difficulties in reading and writing are therefore not confined to children suffering from cerebral palsy, but they appear to be more frequent, more severe and last longer in them, than in those who show no evidence of cerebral abnormality. It is probable that the reason /

stibbars ettt it owl

Run to the tree

su tel ecnad su tel

a ni herd I ihc evi F

This is the way we

dance and sing

12-10-52

Spelling

murd

tab

men

fun

plum

The writing of a female patient with severe spastic tetraplegia but normal intelligence. Aged I^0 at the time of writing the example.

reason for their greater frequency is not to be found in the fact that a high proportion of those with cerebral palsy are mentally defective and therefore less able to overcome their lateralising difficulties than children of normal intelligence. In a recent series of cases of cerebral palsy it was found that the difficulties were more frequent than in a control series of normal children with a similar distribution of intelligence. Dunsdon, 1952. This observer attributed the difference between her groups to the higher proportion of cases with cerebral palsy who showed evidence of mixed hemisphere dominance, as judged by tests for eyedness, handedness and footedness. Unfortunately, her criteria for determining the presence of cerebral dominance must be questioned. It has been demonstrated that children with cerebral palsy do tend to show very defective spatial appreciation in a high proportion of cases. This is so in patients with normal intelligence as well as in those with mental defect, though poor spatial appreciation is commoner in the latter. Lord, 1937. It seems likely that, in most cases, lateralising difficulties with small words and letters should be regarded as one manifestation of disturbance of the appreciation of spatial relationships which occurs in patients with cerebral palsy, but the question must still be asked, whether this disturbance can be regarded purely as a direct result of cerebral injury or whether other factors, /

factors, especially changes of handedness may not also be important causes.

An attempt is being made at present to determine the incidence of significant lateralising difficulty in primary school children in three groups, normal children, mentally defective children and children with cerebral palsy. As a result of questioning a large number of teachers and testing the handedness and intelligence of the children it is hoped to determine the incidence, the severity and the duration of lateralising difficulties in the three groups. It should also be possible to assess to what extent latent sinistrality and crossed laterality is significant in determining the occurrence of lateralising difficulties.

In a small group of children with cerebral palsy who were in normal schools, schools for the physically and mentally handicapped it was possible to obtain the results obtained in table . Much difficulty was found in obtaining information about the early primary school period in the older children and the results cannot be considered as being very reliable. It will be seen that of the 57 patients, about whom it was possible to obtain information, 17, or about one third, showed lateralising difficulties for more than three months. Ten of these patients suffered from hemiplegia. It is interesting to notice that a higher proportion of patients with right hemiplegia /

hemiplegia suffered from lateralising difficulty than of left hemiplegia. Unfortunately the numbers are far too small to be significant but the similarity to the difference in the incidence of aphasia in right and left hemiplegic patients is striking.

Amongst 20 patients in a special school for children with cerebral palsy, none of whom was mentally defective, 5 were found to have had or have lateralising difficulty, four in reading and writing and one in writing alone.

It is not proposed to discuss in detail the nature of the lateralising difficulties and other defects of spatial conception suffered by children with cerebral palsy, nor can accurate figures for their incidence be given. It is certain, however, that they are commoner than is generally realised and it is probable that they are a cause of much educational failure and difficulty.

As more patients were examined during the survey an increasing number were found in whom true aphasia could not be demonstrated, but in whom some degree of word deafness did seem to exist. Unfortunately testing all the children fully enough to demonstrate that such mild degree of word deafness did exist was impossible in the conditions of the survey.

Poor concentration was a marked feature of a high proportion of patients with cerebral palsy. In its extreme /

extreme form it was shown by the hyperactive patients previously referred to; it was present to a less extreme degree, however, in many patients, especially below the age of 9 or 10. In a proportion of cases the defective concentration was accompanied by some overactivity. The education of these children presents a problem which is extremely difficult to manage. In some of the epileptic patients who required heavy doses of anti epileptic drugs to suppress their fits, poor concentration was inevitable.

The education of children with cerebral palsy.

In table LIX are shown the numbers of children of school age in the present series attending different types of schools or classified as ineducable. The figures obtained during the survey are compared to those of other similar surveys. It will be observed that there are differences in the proportion of children attending schools for the physically handicapped and schools for the mentally handicapped in each. It is probable that the major part of these differences may be explained by the differences in policies of the school placing of patients with cerebral palsy, and by variations in the special school facilities available in different places. The proportions of children in normal schools are similar in all three surveys.

It /

Table LIX.

The schooling received by children with cerebral palsy. Percentage distribution of children with cerebral palsy in normal schools and schools for the handicapped.

Survey.	This Survey.	Holoran.	Cohen.
Date.	I953.	I952.	I953.
Number of patients.	I54	I4I	I48
Percentage Normal schools.	26.0	30	35.5
Schools for the physically handicapped.	27.9	47	I7.3
Schools for the mentally handicapped.	24.7	7	47.2
Uneducable.	2I.4	I6	
Totals.	I00	I00	I00

-21-

It seems probable that between one quarter and one third of children suffering from cerebral palsy are capable of benefiting from normal educational methods. The remainder are ineducable or in need of special educational facilities. The proportion of children able to benefit from higher secondary education is very small. Only three patients with cerebral palsy in the present series were thought by their teachers to be capable of attempting the Scottish leaving certificate. One was a patient with dyskinesia and two were patients with hemiplegia. All had intelligence quotients of over 115.

It will be seen from table LVthat over half of the patients in normal schools were cases of hemiplegia. All, therefore, had one functionally useful upper limb. Apart from three, who showed very infrequent epileptic fits of grand mal type and a few who showed letter reversals in writing, associated defects were conspicuously absent. It will be observed that 11 of the patients with diplegia were in normal schools. Seven of these were paraplegic, with functionally useful upper limbs. Only one case of tetraplegic distribution was found in normal schools and his paresis was considered to be of relatively mild severity. Types of cerebral palsy which tended to affect all limbs resulted in a small proportion of children in normal schools. Thus there were only two patients with dyskinesia /

dyskinesia in normal school, though the intellectual impairment was generally less severe in these cases than in patients with other forms of cerebral palsy. Only two patients with ataxia were in normal schools.

Thirty three patients or 21.4% of those of school age in the series were classified as ineducable. In Edinburgh the decision to classify a child as ineducable is based much more on the finding of severe mental defect than on the severity of the paresis for facilities are available for the education of the severely palsied child if his intelligence is adequate. Nevertheless, since severe mental defect occurred in a much higher proportion of patients with severe cerebral palsy than of those who were mildly affected, the majority of ineducable patients also showed severe physical disability.

Thirteen of the patients classified as ineducable were in institutions for the mentally defective, the remainder were in occupation centres or at home. Six had been on institutional waiting lists for more than two years. Eight of the ineducable patients showed overactive behaviour disorder. Twenty-one were bedridden. All were in need of constant supervision and attention.

Eighty-one of the patients of school age, or 52.6%, attended special schools for the physically or mentally handicapped. /

handicapped. The decision as to which school was best for the child had to be decided on the basis of the relative severity of the palsy and the mental defect in the individual case. There was some overlap, therefore, in the intelligence quotients and the severity of the palsy amongst those in schools for the physically handicapped and those in the schools for the mentally handicapped. For the most part, however, patients with moderately severe or severe cerebral palsy and intelligence quotients of over 70 were found in schools for the physically handicapped. Patients with intelligence quotients of over 70 and under 85 who were mildly affected might be found in schools for the mentally handicapped, in those for the physically handicapped, and occasionally in normal schools. Patients with intelligence quotients of less than 70 were found in schools for the mentally handicapped in most cases, whatever the severity of their palsy. A few were in schools for the physically handicapped, however, and two remarkable children, each with an intelligence quotient of between 65 and 70, were found in normal schools. Of the forty-three children in schools for the physically handicapped, six were in residential schools, four in a school for children with cerebral palsy and two in a school for children with physical handicaps. One of the patients in a school for the mentally handicapped was in a residential school.

It will be seen from table LV that the proportions of patients /

patients in schools for the physically handicapped and the mentally handicapped varied somewhat in the different categories of cerebral palsy. Thus approximately half the educable hemiplegic patients were in normal schools, one quarter in schools for the physically handicapped and one quarter in schools for the mentally handicapped. Only one patient with diplegic paralysis of tetraplegic distribution was in normal school. The remaining 6 educable patients were in schools for the mentally handicapped and none was in schools for the physically handicapped. In contrast a much higher proportion of patients with ataxia and dyskinesia were in schools for the physically handicapped than in either normal schools or schools for the mentally handicapped.

The prospects of regular employment for those with cerebral palsy.

The prospects of regular remunerative employment for those with cerebral palsy are poor in present conditions unless their palsy is of relatively slight severity and extent. Referring to patients with diplegia a recent report stated - "The severely paralysed, no matter how "intelligent or capable they may be, do not appear to "stand much chance of finding work in Glasgow." Ferguson, et al. 1952.

Some experience was gained in assessing the employability of patients with cerebral palsy, aged between 16 and /

and 18 years in association with the local Youth Employment Officer in Edinburgh. Unfortunately, it is not possible to give accurate figures for the numbers of employed and unemployed in this age group as the total numbers of patients with cerebral palsy are unknown. Ten patients are known to be employed, however, and seventeen to be unemployed.

On the basis of this experience it was thought to be of interest to attempt a somewhat speculative assessment of the eventual chances of employment of the children of school age in the present series. Patients were placed in three categories, probably employable, possibly employable and unemployable. The results of the assessment are shown in table LX .

The majority of patients capable of attending normal schools were considered to be probably employable. One patient with diplegia of triplegic distribution attending normal school had rather frequent epileptic fits of grand mal type, however, and was moderately severely paralysed. He attended private school and was excluded from the group of probably employable. One patient attending normal school showed tetraplegic paralysis of mild degree but had marked speech and visual defect. He also was excluded from the group of those considered to be probably employable. Two patients with diplegia attending schools for the physically handicapped were felt to be intelligent enough and to have /

TABLE LX.

An assessment of the prospects of employment in present circumstances of 154 children of school age with cerebral palsy

Unemployable		Possibly employable		Probably employable	
Criteria	Numbers of patients	Criteria (All with intelligence quotients above 70.	Numbers of patients	Criteria	Numbers of
Intelligence quotient below 70.		Hemiplegia with epilepsy P.H. schools	5	Hemiplegia. Normal schools	23
Hemiplegia	22	Mild hemiplegia. M.H. schools	2	Hemiplegia. P.H. schools.	3
Diplegia	25	Paraplegia. P.H. schools.	1	Paraplegia. Normal schools	7
Other	15	Triplegia. Normal school	1	P.H. schools.	1
Total I.Q. under 70	62	P.H. "	2	Triplegia. Normal schools	2
Intelligence quotient between 70 & 85 but severely affected		M.H. "	1	P.H. schools.	1
Hemiplegia	3	Tretraplegia. Normal school	1	Others. Normal schools	6
Diplegia	10	Others. P.H. schools	12		
Other	11				
Total severely affected. I.Q. between 70 & 85	24				
Total unemployable	86	Total possibly employable	25	Total probably employable.	43
Percentage	56%	Percentage " "	16%	Percentage probably employable.	28%

have physical defects sufficiently well compensated for to allow their inclusion in the probably employable group. Three patients with hemiplegia attending schools for the physically handicapped were also thought to be probably employable. One of these patients had moderately severe speech defect which was improving and another infrequent epileptic attacks of Jacksonian type. In all, 43 patients or 28% of those of school age were considered to be probably employable.

Patients with intelligence quotients of less than 70 were automatically classified as unemployable. They included all the patients classified as ineducable by the Education Authority. Finding regular employment is a matter of great difficulty for the majority of physically normal adults with intelligence quotients of less than 70. When many of the occupations requiring heavy manual labour are impossible because of physical defect, it is difficult to believe that more than occasional unskilled employment will be possible for any of the patients with cerebral palsy whose intelligence quotients are below this level.

Patients with intelligence quotients between 70 and 85 might be expected to be suitable for unskilled work but if this is rendered impossible by severe physical handicap it seems unlikely that regular employment is possible. 24 patients with severe cerebral palsy and intelligence quotients between 70 and 85 were therefore classified as unemployable. /

unemployable. In all 86 patients were considered as unlikely to obtain regular employment, 56% of those of school age in the series.

Twenty five patients, 16% of the series were classified as possibly employable. Only two cases in normal schools were placed in this category. Twenty patients were from schools for the physically handicapped and included 5 patients with ataxia and 6 with dyskinesia whose intelligence quotients were not less than 70. Three patients from schools for the mentally handicapped were classified as possibly employable. In each case the intelligence was between 70 and 80 and the physical handicap relatively mild.

It seems clear from this rough assessment of the chances of regular employment for the patients with cerebral palsy in the present series that not more than one third are likely to be self supporting. A proportion of the 28% of patients with cerebral palsy who are considered to be employable are likely to obtain only jobs in which the prospects for any advancement are poor.

The 56% of patients in the unemployable group are unlikely to make any contribution to their own maintenance. A high proportion will be in need of constant attendance and nursing care for many years. Of the remaining 16% of patients, in the possibly employable group, it is probable that some could be self supporting, or make a considerable

contribution to their own maintenance, in favourable circumstances. Present conditions, however, are anything but favourable for the severely palsied child in search of employment.

Acknowlegements.

The survey which has been described would have been impossible without the full and ready co-operation of the many clinicans whose patients were examined. Unfortunately they are far too numerous to mention individually but I am especially grateful to the paediatric physicians and surgeons in Edinburgh Hospitals who gave such ready facilities for the examination of their patients, and advice whenever requested.

The officers of the Maternity and Child Welfare Service and the School Medical Service were most generous with their help. Especial thanks are due to Drs. Boog Watson and H.P.Tait for the assistance they gave throughout the survey.

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