EPILEPSY AND CEREBRAL PALSY*

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The two conditions epilepsy and palsy in its many forms are amongst the earliest syndromes recorded in history. The association between epilepsy and cerebral palsy in childhood is a matter of everyday experience, and this association is of scientific interest as throwing light on the nature of cerebral palsy and, more particularly, of epilepsy. It is also of practical importance in view of recent efforts to make more adequate provision for those children with cerebral palsy who are educable. The existence of the two conditions in one child constitutes a double handicap. The present tendency in our educational system is for ever-increasing subdivision of educational 'types', and any child who is difficult to fit into one of the artificial categories thus constructed is in danger of remaining outside the educational system altogether. Epilepsy and cerebral palsy make a difficult administrative problem. Teachers do not much like to have epileptics in their class as they find that the fits are liable to disrupt the work of the class, and epileptic colonies tend to insist on children being able-bodied when admitted for instruction. So the child with both disabilities may be refused admission to one of the few schools for cases of cerebral palsy and also to an epileptic colony. From both the scientific and the practical aspect, then, it is interesting to examine the correlation between these two conditions and to investigate the significance in patients with cerebral palsy of the presence of an epileptic tendency as well. In considering this problem I propose to draw largely on information available about mental defectives, but I believe that what I have to say about these children applies in large measure to others who have not been classed as mentally defective. I have in mind the fact that a child with this double disability is very liable to be classed as mentally defective in any event, irrespective of any further investigation of his mental ability.

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Frequency of Epilepsy as a Complication of Cerebral Palsy

Brissaud and Souques (1904) attempted to confine the term 'Little's disease' to those cases not complicated by fits or mental defect, but Little's (1861-2) own description of 63 cases refers specifically to the complication of convulsions. Kinnier Wilson (1940) refers to Little's disease not as 'an ailment of a well defined character but a mere syndrome and a rather wide-ranging one at that'. Since epilepsy is also not a disease but a symptom of cerebral dysfunction it is understandable that cerebral palsy and epilepsy should often be encountered in the same patient. Kinnier Wilson states that in his experience 30% of cases of cerebral diplegia have fits which may be general or one-sided. He contrasts this figure with that of 60% of cases of infantile cerebral hemiplegia which, according to his out-patient figures, are so complicated. He quotes Gowers, Fuchs, König, Sachs and Peterson as finding a similar proportion affected.

Cerebral palsy comprises a group of conditions of extremely mixed aetiology and morbid anatomy. The incidence of epilepsy in the different groups naturally varies. For example, the infrequency of convulsive seizures in double athetosis is well recognized (Ford, 1944). Any group chosen for study will have a composition depending on the method of selection employed. The figures for the Fountain Hospital (Table 1) are based on some 806 beds for certified mental defectives, most of them children. Cases of cerebral palsy bulk largely among them and in the majority of such cases the cerebral lesion is probably more gross and widespread than in those children with cerebral palsy who are enabled to attend school. To this extent the incidence and type of epilepsy in these patients with cerebral palsy may have a limited general application. It should be mentioned that in this group cerebral palsy occurs also in association with conditions such as microcephaly, hydrocephalus and the like. Nonetheless, there is considerable

overlapping both in respect of intelligence, pathology and incidence of epilepsy between these patients with cerebral palsy in a mental deficiency institution and those treated elsewhere. The figures for this group therefore have general as well as particular interest.

TABLE
PATIENTS IN FOUNTAIN HOSPITAL GROUP

Total number of patients on books (2.12.53)	..	777	
Total number of cases of cerebral palsy (2.12.53)	..	177 (23%)	
Total number of cases of epilepsy (2.12.53)	..	185 (24%)	
Number of cases of cerebral palsy recorded in past 5 yr.	..	228	
Number of cases of cerebral palsy died in past 5 yr.	..	26 (11%)	
Of total cases of cerebral palsy			
Spastic diplegia	.. 103 (45%)	Epilepsy	.. 51 (50%)
Hemiplegia	.. 35 (15%)	"	.. 18 (51%)
Athetosis	.. 19 (8%)	"	.. 4 (26%)
Ataxia	.. 5 (2%)	"	.. 0
Unclassified	.. 66 (29%)	"	.. 30 (45%)
	228		103 (45%)
Number of cases of epilepsy recorded in past 5 yr.	..	265	
Number of cases of epilepsy with cerebral palsy	..	99 (37%)	
Number of cases of epilepsy died	..	49 (18%)	

It will be seen from the figures given in the Table that there is a big overlap in this group of institutional cases between epilepsy and cerebral palsy. Apart from mental defect which is common to the group as a whole, epilepsy is the commonest single complication of cerebral palsy, being present in nearly half the cases. The above figures are compiled on the basis of fits which were actually recorded in the Fountain group of hospitals. Many other children had a history of fits before admission, but these are not considered here. Our experience with this group of epileptics with cerebral palsy shows that they share in common with the so-called idiopathic group of epileptics the tendency to have fits when there is present some additional factor in the shape of intercurrent infection, *e.g.*, measles or respiratory disease. Little significance can be attached to the figures of deaths given since the portion of the five-year period during which the individual patients have been classified as epileptics or cerebral palsied varies considerably.

Fits in Different Forms of Cerebral Palsy

The division of the fits among the different forms of cerebral palsy is interesting. As an arbitrary method of classification the dominating symptom has been taken as a basis though naturally some patients present, for example, both spasticity and athetosis. Roughly half the hemiplegics and half the spastics have fits, which is in contrast to the findings of Kinnier Wilson quoted above. On the other hand the strikingly lower proportion of athetotics so affected, roughly one quarter, is in keeping with the general finding. Only five cases

have been classified as ataxia, but not one of these has fits. These patients are characterized, in addition to the lack of muscular coordination, by a low muscular tone. Among the group of athetotics, those with 'pure' athetosis are the non-epileptics. The fits occur in those athetoid children who exhibit mixed neurological symptoms, *i.e.*, usually including an element of spasticity which persists in between the athetoid movements. In the case of 'true' athetosis, on the other hand, muscle tone is low or even less than normal when the muscles are not actually engaged in an athetoid spasm. The general impression of a correlation between epilepsy and muscle tone is strengthened by observation of those patients in our hospital group who are not cases of cerebral palsy. A particular example is the case of mongolism. These patients seldom suffer from epilepsy and in them muscle tone is consistently and uniformly low. I have reported elsewhere (Kirman, 1951) the exception which proves the rule, the case of a child with mongolism and cerebral palsy, a rare combination, who was also subject to fits. The fits and the cerebral palsy were both due to infarction of the brain associated with a congenital abnormality of the heart.

Where there is a neurological picture which can be described as cerebral palsy, there is an anatomical lesion in the brain, usually obvious macroscopically, and localized to the extent that it particularly affects some part of the motor system, whether it be the frontal cortex or some sub-cortical centre such as the globus pallidus which is commonly damaged in athetotic syndromes. Such lesions are commonly associated with epilepsy. It would not be true, however, to suggest that brain lesions which have no gross macroscopic localization are not also commonly associated with fits. Mongolism is an exception in this respect, but the less common diffuse disorders of the nervous system such as phenylketonuria and the lipid disorders are quite commonly accompanied by epilepsy. Thus, out of the 16 cases of phenylketonuria at present in this hospital group, three are complicated by epilepsy. Of these, one has been classified as a case of cerebral palsy. The other 15 patients are all mobile and have no localized palsies. In the few cases of phenylketonuria which have come to post-mortem examination, there has been little macroscopical change, with the exception of Penrose's patient (1939) who had a presumably coincidental neurofibromatosis. Corsellis (1953) does not report any naked-eye abnormality in the brain of his case, that of a phenylketonuric idiot with epilepsy. From his description the case would not in the ordinary way have been classed as one of cerebral palsy, though

'the lower limbs were slightly spastic and her gait was unsteady. All her voluntary movements were clumsy and uncoordinated'.

A child with lipoidosis is also included in the present series as having cerebral palsy of athetotic type. The issue in this patient was complicated by a history of jaundice of the newborn and by the fact that anti-D substance was present in the mother's blood. However, the brain condition at post-mortem examination and the clinical state were compatible with the picture usually found in amaurotic idiocy. The electroencephalogram supported this view.

Of five cases of gargoylism which have recently been in this hospital, none was handicapped by any specific palsy and only one had fits.

Frequency and Relation to Age

As with uncomplicated epilepsy, fits accompanying cerebral palsy tend to fall off in frequency with increasing age. The total number of fits is also often surprisingly small. In many cases there is a history of only a single convulsion. The relationship between the average incidence of fits per year and age is shown for a section of the group of cases of cerebral palsy in Fig. 1.

This graph has been obtained by adding together the number of fits recorded for a given year of age for all the patients of whom a record had been kept at that year and then averaging. The numbers

of patients observed under the age of 3 or above 19 was too small to justify compiling an average. The low early incidence of fits, with a subsequent steep rise, is probably due to the change in the character of fits as infancy is left behind. The majority of the young infants with an epileptic tendency in our care rarely have fully developed major convulsions, though they may have a variety of other manifestations, varying from attacks of pallor, dreaminess, screaming attacks, to isolated myoclonic jerkings and unexplained falls. Thus the number of fits recorded in infancy may be small. As the nervous system matures the more adult type of fit which is readily recognizable makes its appearance and is recorded on the epileptic chart.

The decline in the number of fits shown at puberty and after is probably compounded of two parts. On the one hand patients with many fits tend to die. Therefore the survivors tend to be those with fewer fits. On the other hand it is my impression that there is also a tendency for fits to become less frequent in certain patients.

Relation of Epilepsy to Intelligence

It will be appreciated that an adequate record of a thing so elusive as an epileptic tendency is at best unreliable. How much more is this true of such a will-o'-the-wisp as intelligence! There is no general agreement as to the nature of this quality and although it tends to be treated mathematically

FIG. 1.—Average number of fits per patient annually in 54 cases of epilepsy and cerebral palsy.

as an entity it almost certainly is not. Tests of intelligence are notoriously subject to error even within the normal range. This is much more the case with a special category of children living in institutions with, in the majority of cases, gross brain abnormality.

Formal testing was done on the majority of the patients with cerebral palsy under discussion and the comparative results for the epileptic and non-epileptic are set out in Fig. 2. The significance attaching to these results is necessarily limited. They are records of performance, usually on several different occasions. The heavy overweighting of the sample of cases of cerebral palsy with low-grade patients makes any attempt at differentiation between epileptics and non-epileptics in point of intelligence difficult. In addition, it is notorious that the application of ordinary tests to patients with cerebral palsy is complicated, since such patients commonly have a disability of both hand and tongue, upon the function of which formal tests are based. After giving due weight to all these considerations it may still be thought that the fact that out of 10 patients with an I.Q. above 50 only one is in the epileptic group is significant. Again, taking the figures for those with an I.Q. above 30 the proportion of non-epileptic to epileptic is 3 to 28, *i.e.*, 10%, whereas the proportion for the whole group is 38% non-epileptic. I am indebted to Professor L. S. Penrose for the following interpretation:

'The correlation between epilepsy and mental grade in cases of cerebral palsy is -0.17 , with a standard error of ± 0.08 . The chances of arriving at this distribution by random sampling would be about 1 in 50.'

Additional figures are necessary to enable definite conclusions to be drawn, but this material does point in the direction that would be anticipated. The fact that epilepsy is present in addition to cerebral palsy would suggest the likelihood of a greater general dysfunction of the brain, likely to result in lower intelligence. In the patients with epilepsy the brain lesions are more likely to be cortical, whereas as has been said, some of the non-epileptic cases have neurological syndromes which suggest that there is only minimal involvement of the cortex. It is quite likely that in some patients the epileptic type of activity in the brain, short of actual convulsions or clear-cut petit mal, may interfere with cerebation to an extent sufficient to impair estimated intelligence. In addition, there is the much discussed possibility that damage to the brain with resulting impairment of intellect may result from the effect of fits. Though it has long been accepted (Meyer, 1939) that such is the case, doubt has been cast upon these observations in some instances (Earle, Baldwin and Penfield, 1953; Falconer, 1953), and the view now gaining ground that a number of the lesions recorded, particularly in the temporal area, are the cause and not the result of the epilepsy.

FIG. 2.—One hundred and sixty-two cases of cerebral palsy showing proportion affected by epilepsy at different levels of performance (I.Q.).

Relation to Electroencephalogram

Bridge (1949) states that out of 742 children attending the epilepsy clinic only 30% showed no clinical evidence of brain damage, as assessed from the history, neurological examination and further investigation. (The possibility exists that a large proportion of these 30% also had anatomical brain lesions.) In the case of epileptic children with cerebral palsy clear evidence of a brain lesion is by definition present and it is sometimes possible to assess the situation and extent of the lesion with some accuracy. For this reason electroencephalographic studies on these cases should be particularly useful as a guide to those cases where there is no such clinical evidence as to the situation of the epileptogenic focus.

It is noteworthy that 'epileptic features' in the electroencephalogram are by no means confined to those patients in the group of cerebral palsy with frank clinical epilepsy. Thus J.A.Y. with microcephaly, spastic diplegia and athetoid movements, classed as an idiot, but with no epilepsy whilst in hospital had 'a doubtful focus of sharp waves on the right in transverse recording'.

Another example of a case of cerebral palsy uncomplicated by epilepsy is that of R.L.R. In his case, however, the electroencephalogram showed 'Frequent bursts of moderate voltage sharp waves . . . in runs of two to four waves at a time. They appear to be bilaterally synchronous and symmetrical and there is no definite cortical focus. The lesion responsible for these abnormalities is probably either diffuse or central and basal.' The other remarkable thing about his record is that it was not strikingly abnormal apart from the above findings, although he is an idiot with an estimated developmental quotient of 4. The record, 'a somewhat low-voltage recording, showed alpha, theta and beta activity in about equal proportions in all areas. There is little change on eye opening and closing.'

In the above case it might be postulated that the abnormal activity reported emanated from some central or basal focus as suggested and so interfered with the function of the potentially normal cerebral tissue, though falling short of clinical epilepsy, as to constitute the basis for the idiocy. In fact, however, the existence of the lowest grade idiocy, without the redeeming feature of the alertness and apparent comprehension which sometimes characterizes hopeless cripples, and coupled with a severe spastic diplegia, leads to the conclusion that there is extensive, generalized brain damage, heavily involving the cortex.

R.R.N., another patient with a history of icterus neonatorum not due to rhesus incompatibility, who

had an athetoid type of palsy showed 'a doubtfully normal record . . . some good alpha can be seen . . . there is no asymmetry and no epileptic abnormalities'. D.W.T., with an athetoid type of palsy associated with toxæmia of pregnancy in the mother, showed 'no cortical focus and the record is doubtfully abnormal for the age'. S.W. was also a case in the athetoid group without epilepsy. Her condition was attributed to rhesus incompatibility but the damage appeared much more severe than usual, resulting in complete idiocy. In her 'alpha activity can be clearly seen . . . this was symmetrical . . . the slow activity present did not appear to be in excess for the age'.

In sharp contrast with the above findings was E.A.C., who, though having a history of icterus neonatorum, presented with a hemiplegia and clinical epilepsy. His record was 'a grossly abnormal record which is almost entirely occupied by a very high-voltage slow spike and wave . . . This activity is bilaterally synchronous and symmetrical . . . There is no evidence of cortical focus'.

The cases of athetosis resulting from rhesus encephalopathy in general had comparatively normal electroencephalograms. Thus, in the case of P.B. 'a clear dominant alpha rhythm at about 10 cycles/sec. could be seen to block with eye opening . . . the record . . . is probably normal', and in J.L.F. 'alpha activity can be seen at 9 cycles/sec. blocking to eye opening . . . No epileptic features and the record is doubtfully normal.' B.L. has a post-icteric encephalopathy which is not associated with rhesus incompatibility. He showed 'an abnormal record in which only traces of theta activity can be seen and the frequency of which is at the upper limit of normality at about 14 cycles/sec. The dominant rhythm appears to be irregular theta activity which is present throughout the record and has no definite focus in the transverse position. No specific epileptic features . . .' A repeat showed 'good alpha . . . which is symmetrical and blocks well . . .'.

Anatomical studies (Crome, publication pending) demonstrate that the only cases in our group in which the cortex has been relatively spared, and where there was a cerebral palsy, were those patients previously mentioned, with a double athetosis. It is not possible to base general final conclusions on the findings in our group of mental defectives but they provide no evidence to support the suggestion of a sub-cortical driving mechanism in epilepsy. Bilateral features can be explained as due to bilateral damage or, in the case of bilateral synchronous activity, as due to the influence of one hemisphere on the other.

Pathological Findings

In 16 cases from the total group of cerebral palsy information about the state of the brain at post-mortem examination was available. The pathological picture varied very widely. A common feature was reduction in the size of the brain. In addition lesions were usually widespread and involved the cerebral cortex, and often other structures as well. The commonest lesion was a diffuse gliosis.

K.A. had a spastic diplegia and was classed as an idiot. The abnormality developed in early life. Fits began at the age of 2. He died, aged 14, of bronchopneumonia. The brain weighed 1,085 g. Almost the whole brain was gliosed with occipital ulegyria and partial atrophy of the corpus callosum. The ventricles were dilated. The cerebellum and the basal ganglia also shared in the destructive process.

S.M.D. also had a spastic diplegia. Birth was difficult but the child was probably abnormal before birth. Convulsions began in the first few days. The child was classed as an idiot. She died, aged 2, of bronchopneumonia and epilepsy. The brain weighed 560 g. and there was external hydrocephalus with ventricular dilatation. There was generalized gliosis with ulegyria, affecting especially the frontal lobes. Coronal section showed a number of cystic areas corresponding to the situation of the ulegyria. The basal ganglia were also affected by cystic degeneration.

W.C. had no major fits at any time, though there was a query as to petit mal in early life. He had a spastic diplegia, was hopelessly crippled, but appeared to have more comprehension than the other patients mentioned. The head was small. Delivery was instrumental with damage to the head. This was thought to be the cause of the mental deficiency and of the cerebral palsy. He died, aged 10, of carbuncle of the kidney. The brain weighed 895 g. There were numerous areas of induration. The ventricles were dilated. The corpus callosum was thinned. Many of the gyri showed destruction of the normal pattern of the cortex. There was also marbling and patchy cell loss in the corpus striatum and thalamus.

The findings of gliotic encephalopathy in the cases referred to above were typical of the group as a whole, both epileptic and non-epileptic. Usually there was gross involvement of both cortical and sub-cortical structures.

Some cases had more specific features allowing of a more precise estimate of the time of onset of the pathological process. Thus F.N. was an epileptic imbecile with a right hemiplegia. She died aged

4 of status epilepticus. The brain weighed 570 g. There was a left hemiatrophy of the brain with localized microgyria more marked on the left side. The presence of this condition implies that the abnormality occurred early in intra-uterine life.

T.J.R. had a cerebral palsy of the athetoid type which constituted a complete bar to all voluntary activity. There were no fits. He had some understanding. He was not jaundiced after birth. He died, aged 11, of whooping cough. The brain weighed 990 g. The pattern of the cerebral gyri was within normal limits and in general the cerebrum appeared normal, though with a suspicion of increased consistency in the occipital lobes. On the other hand the cerebellum was grossly diseased, being small and indented. The cerebellar cortex was gliosed and the dentate nucleus could not be seen. There was an associated pallor of the ventrolateral nucleus of the thalamus.

P.A.N. showed an example of the malformation of the brain likely to occur as a result of a morbid process beginning in early intra-uterine life. Pregnancy and delivery were normal. He had difficulty in feeding throughout. There was a spastic diplegia of extreme degree and he was microcephalic. However, he had at no time had any fits. He seemed to have some comprehension. He died, aged 18 months, of bronchopneumonia. The brain weighed 530 g. and showed arhinencephaly with failure of separation of the frontal lobes, and absence of the corpus callosum.

J.F.K. was a case of epilepsy and cerebral palsy with brain lesions which may have developed at a much later stage. There was no relevant family history. He was the only child and pregnancy was normal. Delivery was one month premature. He developed slowly and at 3 months was troubled by continuous screaming. The head was large, the fontanelle did not close and he had attacks of 'twitching'. He was of idiot level and had an extreme degree of spastic diplegia. He was considered to be blind. He died, aged $3\frac{1}{2}$, of bronchopneumonia. The brain weighed 290 g. There was external and internal hydrocephalus. The junction of the straight and superior sagittal sinus coincided with a thickening which might have been due to an old thrombus. Almost the entire left hemisphere had been converted into a sac. The right hemisphere was also greatly thinned. Most of the sulci were unrecognizable. The cerebellum was small and flattened.

Summary

Children who have both cerebral palsy and epilepsy are liable to be deemed ineducable and

mentally defective, whatever their intellectual potentialities. Epilepsy in association with cerebral palsy is due to a gross brain lesion. This is probably true of many other cases of epilepsy, including the 'idiopathic' variety. Figures for the Fountain Hospital group based on 806 beds are given. These patients are mainly children certified as mentally defective. Two hundred and twenty-eight cases of cerebral palsy and 265 of epilepsy have been recorded in the past five years. Forty-five per cent of the cases of cerebral palsy were also classed as epileptic (fits observed in hospital). The athetotic and ataxic patients had a low incidence of epilepsy. The frequency of fits in epileptic subjects as recorded increased after infancy but declined with puberty. The evidence suggests that cases of cerebral palsy with epilepsy are likely to be less intelligent on the average than those without this complication. The electroencephalographic recordings show that 'epileptic type' phenomena also occur in patients who have not had fits recorded in hospital. As all the patients in this group, except the small minority of athetotic and ataxic patients, had gross cortical lesions, there is no evidence in this material to

support the thesis of sub-cortical epileptic foci. Morbid anatomy showed a wide variety of lesions of which reduction in brain size and gliosis were the most constant. All the brains were very abnormal and most had gross structural abnormalities.

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