THE RÔLE WHICH HEREDITY PLAYS IN INDUCING
EPILEPSY IN CHILDREN SUFFERING FROM
INFANTILE CEREBRAL PALSY¹

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While the diagnosis of infantile cerebral hemiplegia is relatively simple, one may not with the same ease decide whether the child thus affected will superadd an epilepsy in later life. The percentage of liability in which this latter disorder complicates the cerebral palsy of childhood is variously estimated; ranging between 40 and 70 per cent.

What are the factors upon which one may prognosticate any given case will have epilepsy in later life? One ought not to let the matter lie in doubt if there be any more definite criteria to aid. We should not trust to time to give us the solution, inasmuch as epilepsy may not follow the initial palsy for years, even thirty years after the original palsy, during which time the parents and relatives must wait with the greatest anxiety and apprehension. Then, too, we ought to be prepared to take the greatest precaution to ward off a sequential epilepsy if one may be fairly sure it is to occur; the spasmodic state once fully inaugurated in a child affected with cerebral palsy rarely ever ceases. We have known of but one case where this latter result has obtained.

At first thought one might readily conclude that the site and nature of the original insult to the brain would give us sure prognostic signs, but a careful review of the considerable literature of our subject shows that such facts are not of much help, even though one were able to determine the extent and nature of

¹ Read at the annual meeting of the American Neurological Association, Washington, May 7, 1913.

the lesion in a given case of infantile hemiplegia by the clinical symptoms. As a matter of fact epilepsy followed in the most widely variant types of palsy. The severity of the palsy is no sure indication upon which we may depend. Indeed, Freud states that the most intractable epilepsy follows in the wake of infantile hemiplegia when the latter is so little in clinical evidence that one may only postulate its presence by vague and transient exhaustion phenomena which follow the fit. This type constitutes his well-known paradoxical group of "palsy without palsy." Thus it is seen that spasmodic infantile hemiplegia shows the widest and most variant types of palsy. The palsy may range from a complete, disabling hemiplegia or a hemiplegia in which the paretic state is scarcely discernible even by the most rigid tests.

In spite of the fact that many disease processes spread readily in the infant brain, such as foci of inflammation or trauma, one may not rely on this principle in infantile cerebral palsy, inasmuch as the latter disease process is most often not inflammatory in the strict sense, and it is often quiescent when the neurologist is first consulted, undergoing but little or no change in many years. Therefore if the site, extent and nature of the lesion in these cases may not aid us, to what more certain criteria may we turn our attention?

It is known to all that recent heredity studies have forced the conclusion that in the large group of the so-called idiopathic type of genuine epilepsy heredity plays an important if not the most important rôle in the causation. It is present perhaps in seventy per cent. of this type of epilepsy. While this law undoubtedly applies to those individuals in which congenital mental defect obtains independent of the more general deteriorating mental process due to the epilepsy itself, such research is pertinent to our inquiry and thesis, more especially so because a neurotic predisposition also enters to a considerable extent in the causation of cerebral palsy of infancy itself.

We have therefore decided to inquire into a large material of infantile cerebral palsy and compare the heredity rôle in causation of the palsies plus epilepsy with that seen in the so-called genuine disease. We are aware, of course, that many are not wanting who hold that infantile cerebral palsy and epilepsy are but two expressions of the same disease process in the developing

fetal and child life. If an infantile cerebral paralytic may be made epileptic by the interposition of a slight trauma or the perturbing influence of puberty—and most frequently no cause for the first fit is brought forward in the history of spasmodic infantile cerebral palsy—how much more potent in causation should be the presence of a spasmophilic or simple neurotic heredity in influencing or deflecting the function of a brain already crippled. The importance of an inquiry of this sort has a distinct bearing upon genuine epilepsy, as it has been shown that 35 per cent. of all epileptics show a preponderance of one-sided convulsions, thus indicating the possible dependence of such a disorder upon a cerebral lesion. Further, more than 12 per cent. of all epilepsies follow in the wake of well marked cerebral palsies of infancy. This proportion obtains in the admissions at the Craig Colony for Epileptics, at which institution our inquiry was made. In our heredity study we were blocked at the outset from making the issue final, owing to the inability of direct comparison of a large material of palsy cases *with* and *without* epilepsy. A great number of palsies without epilepsy were nowhere obtainable, although all the large clinics of New York were laid under contribution, from which fact one might legitimately infer that the percentage of palsy without epilepsy is even greater than that ordinarily held. In the small material drawn from private sources and clinics, in which a sequential epilepsy could be absolutely excluded, a history of a spasmophilic and neurotic constitution was negligible. We had to content ourselves, therefore, with comparing the heredity history of infantile cerebral palsy associated with epilepsy with that obtainable in the so-called idiopathic cases. The data under investigation consisted of 443 cases of palsy with epilepsy, and 443 suffering from the genuine disorder. This material for convenience of comparison is tabulated and charted, and shows in general terms that in the 443 hemiplegic epileptics in the first 3,500 cases admitted to the Craig Colony there were 133 in which no record of hereditary influences in the ancestry could be obtained, which is a little less than one third. In the remaining 310 cases, or 70 per cent., a definite record was found of hereditary influences and the most important of these bearing on the present study are shown in the chart. Epilepsy and convulsions occurred in 115 instances, or approximately 25 per cent., and in a number of cases several of the ancestors of a given

patient were afflicted; alcoholism was mentioned 110 times; insanity 46 times; idiocy and feeble-mindedness in 10 instances; fainting spells in 17 instances; hysteria, neurotic constitution, migraine, etc., in 100 instances; and apoplexy and paralysis in 38 instances.

While tuberculosis, rheumatism, cancer and other constitutional diseases may not be of importance in determining epilepsy, these disorders have been tabulated to show the frequency with which they were mentioned in the ancestry and which might be a factor in producing a lowered resistance in the patient,—tuberculosis was mentioned in 97 of the ancestors and rheumatism in 76.

An equal number of epileptics without hemiplegia have been tabulated. The remarkable similarity is at once manifest. The amount of known heredity is about the same in the two disorders. Alcoholism is a little more frequent in idiopathic epilepsy while spasmophilia occurs in but 17 cases more than in hemiplegic epilepsy. Then, too, tuberculosis is greatly in excess in the idiopathic epilepsy group. Otherwise the comparisons are not essentially dissimilar.

But few data of other observers are at hand for checking up our study. Bratz, in comparing a small material of less than 50 cases, states that the heredity in idiopathic epilepsy and hemiplegic epilepsy is not dissimilar, and his study tallies with ours that there is no essential qualitative or quantitative difference between the heredity in the two conditions. He mentions no rôle the heredity may play in exciting an infantile hemiplegia to take on a sequential epilepsy.

Fuchs's study based upon an analysis of but 100 cases showed some sort of heredity element in only 29 per cent. of the palsy material, while in the 44 cases which later became actually epileptic an heredity history was present in 34 per cent. Thus the lesion itself, if one may be permitted to argue in this manner, played but one third of the rôle in causation. In other words, the ratio of heredity to palsy was about 1 to 4, while heredity to hemiplegic epilepsy was 3 to 4.

The fact that epilepsy usually occurs much earlier in life in hemiplegic epilepsy than in the idiopathic type may be counted both for and against out contention in this paper. Yet it is frequently seen, as mentioned before, that hemiplegic epilepsy occurs

CHART No. I. Comparison of the cases of Epilepsy with Hemiplegia, indicated by the heavy upper line, with an equal number of cases of Epilepsy without Hemiplegia, indicated by the lighter, lower line.

years after the original cerebral trauma and wherein no new palsy symptoms have been shown to occur, as far as can be observed, to account for the added fits. Again, genuine seizures are far from rare in hemiplegic epileptics. Even absences and psychic attacks are not wanting. Usually the same factors, such as indigestion, fright, etc., are equally alleged to have caused both types of convulsions. One knows to what degree the alleged excitants can be considered as the true cause of the genuine disorder, and we presume to class the initial palsy and the sequential alleged causes together and place our greatest reliance upon the absence or presence of an heredity factor in the specific case and prognosticate by that chiefly if not solely.

From the material which we have here submitted, we believe we are justified in concluding that the heredity factor in hemiplegic epilepsy is only a little less in evidence than that of genuine epilepsy, and that the prognosis as to a sequential epilepsy following in the wake of a given case of infantile cerebral palsy should be based upon the presence and degree of spasmophilic or simple neurotic history, and that this latter fact should be given as much or even more weight than the site, nature and degree of the initial cerebral injury as expressed in the palsy.

Our best thanks are due to Dr. William P. Shanahan, Superintendent of the Craig Colony, for permission to use the extensive clinical data and case records at the Colony on which material of this study has been made.