Section of Neurology.

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Pyknolepsy : A form of Epilepsy in Children, with a good Prognosis.

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ABOUT two years ago, a mother brought her little boy to me because he was suffering from "nervous attacks" which the doctor in the country called epilepsy; these attacks, she said, began suddenly about two years before without apparent cause, and had recurred daily with great frequency—"dozens every day"—ever since; this in spite of prolonged treatment with bromides, luminal and other remedies that had been ordered by various consultants she had seen. Apart from the attacks the boy was perfectly well; he learnt his lessons with ease and was good tempered, intelligent, and everything that a healthy boy of his years should be.

After a long conversation with the mother I saw the boy and during the short time he was in my room he had three seizures, all of them exactly alike and, according to the mother, exactly the same as all the others he had had. Whilst sitting in a chair his limbs relaxed, he stared vacantly before him, the eyeballs rolled upwards, the lids flickered once or twice and then after a few seconds he looked about him, smiled, and was himself again.

Everything that I had heard and seen seemed to confirm the diagnosis of epilepsy, but several features of the case struck me as being rather unusual—in particular the great frequency of the attacks from the beginning, their sameness, their uniform mildness and their obvious harmlessness.

Could this be the pyknolepsy of which I had read? Without any feeling of strong conviction in my own mind but desiring to afford the mother some crumb of comfort I ventured to tell her that it was just possible that the attacks would cease some day as suddenly as they had begun.

Nine months later I met the family doctor who asked me if it was true that I had told Mrs. X. that her son would have no more attacks. I hastened to defend myself but he cut short my protestations by informing me that the attacks had indeed ceased a few days after the visit to me and had never recurred. I heard a day or two ago that the boy is still perfectly well.

This may or may not have been a case of pyknolepsy, but after this surprising experience I set to work to learn all I could about this remarkable disease, which resembles epilepsy very closely in its cardinal manifestation yet differs from it most happily in its prognosis, and since that time I have kept a sharp look-out for similar cases—not without success. But my own cases are few and the period during which I have had them under observation is too short to allow me to formulate any conclusions that are likely to convince you.

My remarks this evening, therefore, are based upon a study of cases that have been described during the last seventeen years in foreign journals devoted to the study of neurology and children's diseases, in particular of those cases in which the period of observation has been a long one. Nothing has been written upon this subject in English, so far as I know, and it is because the available information is widely scattered and because I think the time has come when we should define our attitude in the matter, that I bring it before you to-night.

The first descriptions of the disease were given in 1906, by Friedmann, in a paper entitled "On Non-epileptic Absences or Short Narcoleptic Attacks," and in 1907, by Heilbronner, in a paper on "Gehäufte kleine Anfälle" (frequent, or better, aggregated minor attacks). In 1912 Friedmann returned to the subject and after expressing surprise that his first paper had given rise to so little comment he described some fresh cases, and gave a highly favourable account of the after-history of the patients on whom his original description was based. He should be more satisfied now, because the disease is often called Friedmann's disease, or the Friedmann-Heilbronner disease. Special papers have been written upon it by Mann, Engelhard, Sauer, Schröder, Redlich, Bolten, Cohn, Oppenheim, Stier and others. It is mentioned in every modern discussion on epilepsy and has even received consideration in the latest editions of the hand-books of Kraepelin, Oppenheim, Ziehen, and Lewandowsky.

According to Friedmann the disease has one symptom and one only—the attacks. These begin suddenly in healthy children between the ages of four and ten years, and recur with great frequency—from six to one hundred daily —in spite of well-planned treatment by bromides and other remedies. Ultimately, after persisting for periods which vary from a few weeks to many years they cease, often suddenly, sometimes gradually, and leave no trace of their former presence. Apart from the attacks the child is perfectly well and its physical and mental development is unaffected. The attacks themselves are remarkable for the monotony of their course and their uniform mildness. They consist in an inhibition of the higher psychical processes lasting from five to ten seconds. The power of speech and of voluntary movement is in abeyance, but automatic movements are retained; the child sits or stands with the limbs relaxed, staring vacantly before him, the eyeballs may roll upwards, the lids may flicker but there are no convulsive movements and consciousness is never entirely lost. After the attack the child is well at once and continues his interrupted game or task as if nothing had happened. The attack by itself, as you see, is indistinguishable from ordinary *petit mal.*

Now according to prevailing notions the prognosis in cases agreeing with the description I have just given should be bad, for the fits begin in early life, they are very frequent and they are uninfluenced by the ordinary remedies; yet, in Friedmann's second paper giving the after-history of the patients on whom this description was based, we find that the attacks, after persisting for five years in two cases, and for seven and nine years in others, ultimately ceased and remained absent for four and a half years, for five years, for nine years and for ten years in different cases.

One of Heilbronner's patients was still well six years after the attacks had ceased. One of Husler's patients was still free after three years; Stier's patients, who recovered after having attacks for 2½, 3, 3, 3½, 6, 6½ and 8 years were still free after 2, 3½, 4, 5, 5, 6 and 8 years and others have been described in which the period of freedom from attacks, and from treatment of course, had already extended over from one to three years. One of my own patients, who had frequent

attacks for two years, has been free for eighteen months, another who had attacks—every few minutes as the mother said—for six months has been well for two years and five months, another in whom they persisted for three years has had none for one year and four months, and I have others under observation in whom the period of freedom is still a short one or in whom the attacks still occur. To my own cases I attach very little importance. They interest me greatly, but I cannot expect you to think much of them for three reasons: (1) The period of observation is short; (2) the period of freedom from attacks is short, and (3) the patients are still young. But these objections will not hold against the cases I have culled from the literature, for in these the period of observation has extended to as many as sixteen years, the period of freedom from attacks has been as long as ten years, and the patients have reached such ages, taking a number of cases at random, as 14, 16, 18, 18, 20 and 22 years. These patients were perfectly well, mentally and physically, at the time the final observations were made and although the attacks had occurred daily for many years none of them ever had a single major attack. Do you think that these patients were suffering from epilepsy? This depends, you may say, on what I mean by epilepsy, so perhaps I should ask, were these patients suffering from the epilepsy of the text-books? Surely not. In epilepsy, according to Kraepelin, spontaneous recovery does not occur in more than 2 or 3 per cent. of cases. Gowers, writing on the prognosis with regard to a spontaneous termination, says that this is an event too rare to be reasonably anticipated in any given case, and our President (Dr. Collier) has expressed a similar opinion. In these cases, and in all the others that comply with the strict requirements that I shall mention later, spontaneous termination is a rule to which I have been unable to find a single exception. In epilepsy the prognosis is bad in cases with frequent minor seizures and in cases that do not respond favourably to appropriate remedies. In these cases the attacks are very frequent and the ordinary remedies have no appreciable effect upon them. In epilepsy slight minor attacks rarely attain a maximum frequency at the onset, or recur alone with absolutely uniform severity for more than a short time; almost from the beginning succeeding attacks vary in severity and the first major attack is rarely delayed for more than two or three years. In these cases there is no gradual onset, no climax; the very acme of the disease is attained at once and its most characteristic feature is the monotony and the uniform severity of minor attacks that may recur daily for many years without the occurrence of a single convulsive seizure. In the one disease, mental deterioration and psychical defects are characteristic, almost constant —some would say inevitable—sequels, that appear soonest and most often in its victims who have frequent abortive seizures: the other, in spite of its long duration and the frequency of the attacks, does not impede mental development nor give rise to psychical defects. These—I need not mention others—are differences that cannot well be ignored.

You may not agree with Friedmann that the cases that he and others have described are examples of a disease that has nothing whatever to with epilepsy, but I think you will agree that they deserve a distinctive name. Quoting Sir Clifford Allbutt:

"The right question is this: have I noted in a moving equilibrium, say in Man, that a certain series of changes, static and dynamic, has occurred more than once? If so, was the recurrence still fortuitous, or was the series a case of an orderly recurrent mode which hitherto had escaped attention? If so, the recurrence will be observed again and again by myself or by others in approximate uniformity. The cases of the

newly observed series will vary, some indeed so far as to trespass upon other nosological series already recognized, and no strict demarcation can be drawn around them; yet there may, nevertheless, be difference enough and constancy enough to make it worth our while, for the convenience of observation and thought, to erect the new series into a category of its own, and to stick a label on it?" [1]

I think these cases are worthy of a label, and as the choice seems to lie between non-epileptic absences, short narcoleptic attacks, aggregated minor attacks, and other names equally awkward, I propose that the name pyknolepsy, suggested by Schröder, be adopted by English observers. It is concise, it is non-committal; it refers to the main feature of the disease—the heaped-up, closely packed, aggregated attacks—and it allows us to coin a handy adjective, "pyknoleptic," by analogy with epileptic. Whether you regard pyknolepsy as a symptom, a disease, a morbid entity or what not, is after all merely a matter of taste. Following my guide in these matters again, "by disease I mean a mental concept of a series of symptoms recurring with fair uniformity," and for me pyknolepsy is a disease.

Having thus burned my boats, I propose to discuss the disease in slightly greater detail, because this bears directly on the questions to which you will certainly demand an answer, namely, "Is the prognosis good in *all* the cases that conform to my description, and is it possible to detect the cases with a good prognosis at or soon after the onset of the attacks?" I have arrived at my answers after a careful comparison of the features of the cases that recovered with those in which somewhat similar frequent minor attacks occurred at the onset of ordinary epilepsy.

(1) *The Age of the Patient at the Time of Onset.*—According to Friedmann, the onset in favourable cases occurs between the ages of four and ten years. Subsequent observations, in particular those of Stier, have shown that it is always between the fourth and twelfth years, whereas it is often above or below these ages in cases that resemble pyknolepsy for a time and yet prove to be epileptic. I do not doubt that favourable cases with an earlier or later onset will be encountered in the future, but for the present we must be bound by these limits.

(2) *The Duration of the Disease.*—This varied in cured cases from seven weeks to nine years with an average of about three years. A long duration, therefore, is consistent with a good prognosis. On the other hand, minor attacks of sorts have been known to occur alone in true epilepsy for as long as seven or eight years, hence a long duration of attacks classifiable as minor, but differing in the direction of greater severity from the attacks of pyknolepsy, does not justify a good prognosis. No case has been recorded of frequent minor attacks such as I have described, beginning in childhood and persisting alone beyond puberty, in which epilepsy has ultimately supervened. Once puberty is safely passed the ultimate prognosis, so far as we know at present, is always good in spite of persisting attacks (Stier).

(3) *The Mode of Onset, Frequency, Course and Mode of Cessation of the Attacks.*—In the favourable cases the onset is almost always sudden—"explosive" as Friedmann said, the attacks occurring with maximum frequency from the first day onwards; but in rare instances the minor attacks of true epilepsy also begin in this way; so an explosive onset, though almost constant in pyknolepsy and highly suggestive of it, is not in itself decisive in the differential diagnosis from epilepsy. The frequency of the attacks varies

[1] "Notes on the Composition of Scientific Papers," 1905.

greatly in different cases, from six to a hundred in a day, but is often remarkably constant in each case. Friedmann attached particular importance to this steadiness of course, because the attacks in his cases were repeated daily at about the same rate; but it has become clear that a waxing and waning in the daily number occur in favourable cases, and that changes in frequency from time to time are consistent with a good prognosis. The termination of the disease is usually sudden, but a gradual diminution in the number of the attacks and temporary periods of freedom have preceded their final complete disappearance in several instances.

With regard to the separate attacks: their great distinguishing feature is their extraordinary monotony; in each patient all the attacks are almost exactly the same in their course and severity. In my opinion this is their most important single characteristic, and, by using it as a test as I read the descriptions of series of cases, I have been able in every instance to foretell the sequel and to detect the cases that resembled pyknolepsy in some respects for a time but proved eventually to be cases of epilepsy. The attack is always short, a second or two up to ten or twenty seconds, and very mild. The limbs relax, but the child rarely falls, objects are rarely dropped from the hands, the colour does not change appreciably, there is no mental alarm or physical distress. At most the eyeballs may roll upwards, the lids may flicker, the head may be turned to one side or the arms raised by a feeble tonic spasm, but chewing or swallowing movements or clonic spasms of any kind never occur. At the end of the attack the child behaves as if nothing had happened, and is never languid or sleepy or confused for more than a moment or two.

I have said that bromides and luminal have no appreciable effect on the attacks. This has proved so constant that a favourable prognosis cannot be entertained in a case that responds favourably to these remedies.

In favourable cases it is rare to obtain the family history of alcoholism, epilepsy or nervous disease that is so common in epilepsy, but the children themselves may have suffered from spasmophilic symptoms, or from so-called occasional convulsions, for example at the onset of an acute disease or during an attack of whooping-cough, and a history of these does not prejudice the prognosis.

These are the facts. Now my study of a large number of cases with frequent minor attacks has taught me *first* that *all* the cases with a good prognosis have certain features in common; and secondly—(this is equally important for my argument)—no case with an unfavourable outcome presented all these common features. It follows that if a given case presents these common features the prognosis is good, that is to say it is one of pyknolepsy, for this, by definition, is a form of epilepsy in which the prognosis *is* good. An enumeration of these features, then, should serve as a basis for a short definition of the disease, a definition which will embody a number of requirements that must be met before the diagnosis of pyknolepsy can be made with certainty. I define pyknolepsy as *a disease with an explosive onset, between the ages of four and twelve years, of short, very slight, monotonous minor epileptiform seizures of uniform severity, which persist for weeks, months or years, uninfluenced by anti-epileptic remedies, without impeding normal mental and physical development and ultimately cease spontaneously, never to recur.*

Clonic movements, however slight, obvious vasomotor disturbances, mental distress, palpitations, and lassitude or confusion after the attack, are equivocal symptoms strongly suggestive of epilepsy, and for the present they should be considered as foreign to the disease.

Now to answer my questions: First, is the prognosis good in all the cases that conform to my description? Yes, so far as I know at present. I have read of cases of epilepsy with an explosive onset, of cases in which frequent minor attacks persisted alone for many years, of cases of undoubted epilepsy of long duration without mental changes of any kind, of cases that did not respond to the usual remedies, of cases, in short, that resembled pyknolepsy in some respects; but I know of no case that satisfied all the requirements that I have just laid down and proved nevertheless to be one of epilepsy.

To the second question—Can these cases be detected at the onset? the answer must be, No. There is nothing in the attacks by themselves to distinguish them from epilepsy, and a certain period of observation is necessary before a final decision can be made. In practice it will often happen that the disease has been present for a year or more when the patient first comes under observation, and a careful consideration of the history will give decisive information.

There is another highly practical question that I have not yet considered. It is one that will confront you very often if you study your cases of minor epilepsy closely, as I have done during the last two years, with a view to detecting cases that belong to the category we are now considering. It is this: what is the prognosis in cases that deviate, in the direction of slightly greater severity, from the description that I have given? when, for example, the onset is not explosive or the child falls in some of the attacks, or allows objects to drop from the hands, or changes colour or wets himself sometimes, or is confused for a short time after some of the attacks. All I can say now is that occasional falls have occurred in favourable cases—perhaps the child was in an attitude that rendered a fall inevitable when the attack came on, that objects have dropped from the hands—this, too, is easy to understand, that involuntary micturition did occur in one or two favourable cases; but these symptoms, though undoubtedly consistent with a good prognosis, were more often the first warnings of oncoming epilepsy, and must be looked upon with great suspicion. Until we know much more about pyknolepsy, it is wiser, I think, to include under this name only the cases that conform strictly to the description I have given, otherwise all sorts of frequent minor attacks will be called pyknolepsy, and the great advantage that is gained by limiting this name to those in which the prognosis is certainly good will be lost.

On the nature of the disease I have little to say. Friedmann thinks that it is a form of narcolepsy, but it has very little resemblance to the original narcolepsy of Gélineau. Heilbronner discussed the problem as if the alternative was epilepsy or hysteria, but we know now, that various seizures occur in children that have nothing to do with either. In several cases the attacks began after an emotional disturbance, but in the great majority they began without apparent cause. There is nothing hysterical in the ordinary sense about these young people, and I know of no form of hysteria in which one manifestation alone is present throughout long years. Mann found increased excitability of the peripheral nerves to electrical stimulation in two cases, and concluded that the disease was connected in some way with spasmophilia, but this is certainly not true. Nor is there any reason to believe that the disease is confined to children who possess the so-called psychopathic, neuropathic or vasoneurotic constitutions. Epilepsy alone remains. I shall not repeat my reasons for separating these cases from epilepsy, but if you think of all the conditions that have been included under the heading epilepsy or epileptic equivalents, perhaps you will agree that there is something to be said for a move

in the opposite direction; and when you consider the effect upon the relatives when the diagnosis of epilepsy is made and the utter hopelessness of this dread disease when it comes on in childhood, perhaps you will agree that the cases I have described are worthy of a distinctive name.

May I remind you, in closing, of the long periods of freedom from attacks in some of these cases, up to eight, nine, and ten years, the long periods of close observation by competent observers, up to sixteen years, and the relatively advanced—shall I say safe?—ages that have been attained by some of these patients, up to eighteen, twenty and twenty-two years—patients who suffered for years from the attacks that I have described and are now perfectly well in every respect.

The question whether pyknolepsy is a disease *sui generis* does not concern me greatly. It has been answered in the affirmative abroad, almost without a dissentient voice. I leave it for discussion. For myself I shall be well satisfied if I have succeeded in making it appear probable to you that there does exist a form of epilepsy in children in which the prognosis is good.