THE POST-GRADUATE MEDICAL JOURNAL

VOL. IV.

JUNE, 1929.

No. 45.

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THE DIAGNOSIS OF CHOREA.

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TYPICAL chorea is easily diagnosed by any physician who has previously seen a case. Not all cases, however, are typical ; and owing to the seriousness of the rheumatic heart disease which is part of the picture of true chorea it is of the utmost importance for the physician to be able to diagnose the slight or atypical cases. It is equally important for him to be able to separate off the cases resembling chorea, but not truly choreic, so that he may not treat by

prolonged rest in bed a child no more liable to heart disease than a healthy child. When a patient is being examined, and the possibility of chorea has arisen, the diagnosis is made clear by a consideration of what may be called positive and negative factors. On the one hand are the symptoms and signs of cases of proved chorea. On the other hand are the symptoms and signs which characterize the various non-choreic forms of fidgetiness, and which by their presence exclude chorea unless two unrelated disease processes are to be diagnosed in one case. Before describing these positive and negative factors it is necessary to note certain very much less common central nervous system diseases which may give rise to difficulty. For instance, a certain proportion of cases

of epidemic encephalitis start with movements indistinguishable from those of chorea, usually severe, and only diagnosed by the course of the disease. In an epidemic the physician is on the look-out for this type of case, and by early lumbar puncture makes the diagnosis certain. When other signs of encephalitis are present at the onset less difficulty is experienced, though with a very restless child with strong choreic movements and great emotional lack of control the practical difficulty of recognizing slight cranial nerve paresis may be immense.

Occasionally, too, the athetoid movements of an infantile hemiplegia may lead to mistaken diagnosis, but not if the history of the case be carefully taken, and if the patient be examined.

But this article deals principally with the difficulties experienced in everyday practice, which amounts to the separation of chorea, a disease probably of infective origin and part of the acute rheumatism of childhood, from the abnormal movements of emotional origin, not associated with heart disease.

POSITIVE FACTORS.

The typical case of chorea may be described as follows:—

A girl, aged 8, has been healthy all her life until this summer. She is one of a family in which a sister or brother has had acute rheumatism or chorea, and, perhaps, rheumatic heart disease.

The patient has very likely been a normal happy child, not specially excitable or nervous, playing normally with friends, and eating well. Up to the onset of the illness her sleep was good, not restless. She did not wake with night terrors. Bowels normal, micturition natural; no increased urgency or frequency.

This summer she has been getting tired, and has had occasional pains in the ankles, wrists, shoulders.

Three weeks ago she started abnormal movements, perhaps after a fright, and became emotionally unstable, liable to cry

and laugh, changing moods rapidly. She has also been weak down one side, so that she avoids using the affected arm, and limps a little.

Sleep is fairly good and the movements nearly cease in sleep. There has perhaps been some occasional increased urgency of micturition, part of the general incoordination. For a week she has developed a difficulty in speaking. As she begins to speak she splutters out too much at once and gives it up when the wrong sounds come. This obviously distresses her very much.

As you go to examine her you find her movements increase. You watch and see no movement repeated, but you witness a continuous series of muscle or muscle fibre contractions completely without design or order. The speed of the movements is moderate, less than that of the repeated movements of tics, and yet greater than the athetoid movements of the congenital hemiplegia.

The movements may be almost entirely confined to one side, but usually the whole body is affected, those of one limb or side being exaggerated.

The head and face are affected and the sudden snapping in of the tongue after a momentary delay is very characteristic.

It may be obvious that there is paresis of a limb, and yet in the case of an upper limb the grip on that side may be almost as good as that of the other hand. The tendon reflexes may be normal or increased. In a severe case the chorea knee-jerk may be present, with delay in relaxing.

Absence of all other signs of organic disease of the central nervous system is the rule. The only other signs besides the movements and paresis and emotional instability are those of associated rheumatic heart disease and of acute rheumatism, when such are present. The heart disease may be actual carditis (shown in dilatation, increased rate, weak first sound at the apex, pericardial rub), or evidence of old heart disease (hypertrophy, established mitral or aortic disease, pericardial adhesion), or both.

The presence of any definite rheumatic sign practically clinches the diagnosis.

On the other hand, in any one case absence of carditis is not an argument against a diagnosis of true (rheumatic) chorea.

The presence of paresis is of great help in diagnosis. The paresis and the movements are often most marked on the same side, in the same limb. I have known a doctor diagnose hysterical paresis of the lower limbs as a separate disease in a child with active chorea. According to my experience hysterical paresis is rare before puberty, very much less common than chorea. It is my practice to use the presence of paresis in a doubtful case as an important argument in favour of true chorea.

Moreover, I do not consider the paresis is a conversion-hysteria paresis brought out by the emotional instability that is part of the chorea picture. It seems much more likely that the paresis and the movements are both due to a rheumatic encephalitis and independent of previous emotional disorder. The localization of the paresis tends to be constant in any one patient, while paresis exists.

To illustrate chorea, then, I have taken a child who has not been excitable or nervous up to the time of onset of chorea. Apart from recent symptoms suggestive of sub-acute rheumatism, the child has been in good health. The illness is characterized by three symptoms: Movements, paresis, emotional instability.

The emotional instability is not like the excitability or the nervousness (about to be described) of the two chief types of non-chorea case. There is no particular direction of alteration of emotional type, but it seems that the normal reactions appear more suddenly and cause confusion. Between the attacks the mental state tends to return to normal if the child is of normal emotional development.

NEGATIVE FACTORS.

Owing to the present campaign, whose object it is to find early heart disease and

to prevent heart disease in children, a large number of children are being sent to physicians by parents, teachers, or school medical officers, as "possibly choreic." A few of these children certainly have chorea, and a considerable number certainly have not chorea. The diagnosis of the difficult cases is helped by a consideration both of the characteristics of true chorea (positive factors) and of the characteristics of the types sent as ?-chorea which are certainly not chorea (negative factors).

For brevity and clearness the cases sent as ?-chorea, but which are certainly not chorea, will be described under three heads, though the three types overlap and inter-mingle.

(a) *Tics, Habit-spasms*.—These names are used to designate repeated movements. It is as if an electric current had been applied to a group of nerve fibres, causing a corresponding group of muscles to contract again and again. Yet careful observation shows that the contraction is purposeful, cortical. In fact, one explanation of these movements which is given is that there is always at the start a definite cause, such as a rough collar, a long forelock, an error of refraction (in the case of blinking), tight braces, a coat fitting tightly under the armpit, and so on. These causes, however, do not explain the *persistence* of the particular movement or the *replacement* of one tic by another in one patient.

The movement is evidence of a lesser or greater disturbance in emotional development. It seems possible clinically to distinguish between two kinds, one associated with a rather obstinate disposition, and the other associated with the fidgetiness (*b*) to be described immediately.

These movements have no relation to heart disease. They are very common, and treatment is difficult but often unnecessary.

The occurrence of an attack of true chorea in a child with habit movements does not, in my experience, make the habit movements either disappear or increase.

(b) *Fidgetiness, Excitability*.—The word

excitable is used here as a complement to the word nervous. Excitability and nervousness do not go together so often as is sometimes expected by people who do not really separate the two terms in their minds.

The typical child of this group is excitable but not nervous. Boys are at least as often affected as girls.

The history is likely to be as follows:—

The boy of 6 is brought because of fidgetiness. He is a healthy boy, but has always been very excitable. Lately this may have become worse. He is very lively and never still. He is thin but wiry. At meal times he will not sit down, but must be about the room. He may be fidgety in school, but sometimes he may be quiet at school though fidgety at home. His sleep is restless but he continues to sleep, not waking with night terrors. He hates to go to bed, and is up with the lark in the morning.

His fidgetiness is of the type that might make anyone say, "Do you want to go to the lavatory?" And this leads to the cardinal symptom of the illness, by which anyone may recognize the typical case—occasional or permanent increased urgency or frequency (or both) of micturition, an expression of an abnormal sensitiveness of the urinary tract. Occasionally, in girls, the complaint is of a burning sensation on micturition. This has no direct relation to the common form of nocturnal enuresis, though there may be occasional enuresis due simply to increased urgency. At night the child seldom has to get out of bed to micturate.

This micturition disturbance is frequently found in this type of case, and it is so seldom found apart from the fidgetiness, except in urinary tract infection, that great reliance may be placed on it. That is to say, fidgetiness of this type, present in a child as a "part of his or her nature," is a disturbance of emotional development, is not chorea, and is not associated with heart disease.

This form of fidgetiness often does not closely resemble chorea; but, on the other hand, in certain cases, the diagnosis from

inspection only may be extremely difficult. Hence the importance of care in taking the history of the case.

(c) *Slight Fidgetiness, Chronic Nervousness.*—The child who belongs to this group is not typically excitable, but is chronically nervous, scared. This form of illness, like chorea, is rather more common in girls than in boys.

This child does not look like a severe chorea patient, but the slight movements and emotional state remind the physician of chorea, and especially so because of the vasomotor instability which goes with the state of chronic fear. This gives a complexion like that of subacute rheumatism. Indeed, the diagnosis in these cases is often very difficult.

However, there is this definite type of child commonly sent to the physician as ? - chorea, and who yet has not chorea, and an attempt must be made to avoid treating as a potential heart case any child whose heart is healthy, and especially one who suffers from chronic nervousness.

The patient this time is a girl or boy brought to the doctor by parents or teacher because of tiredness, aching pains, or for nervousness or night terrors.

This patient may be an only child, whose parents are particularly "nice," and are careful with whom the child mixes. Or she is a child who is one of several in a family, but who stands out from the others in being much more nervous.

Although there may have been a recent increase in the symptoms, in this typical case the parents give the history that the child is "by nature nervous." This corresponds with the history of the excitable, fidgety child (b), and often distinguishes both from typical chorea. For in chorea there is generally, in my experience, a history of onset of illness.

Routine questions elicit the information that micturition is normal, but that sleep, though not restless, is disturbed. Frequently the child wakes up suddenly, terribly scared.

This may be twice a night or only a few times a week. In the day the child may be scared of the dark, strangers, a knock on the door, being left alone, traffic, and so on, or may just seem chronically frightened, apart from external cause.

On examination of the child—who is often sensitive, tall, and not thin—the physician notices a faintly suffused pallor which reminds him of the complexion of subacute rheumatism, and which is due to the vasomotor condition, the result of chronic anxiety. This vasomotor state is often reflected in skin rashes, patches of erythema, frequently with an urticarial centre. These patches may appear spontaneously, like blushes, under the physician's gaze, or may appear where the stethoscope or clothes have caused pressure.

The movements are just those which we popularly associate with apprehensiveness and anxiety. A good shot at Bisley is not of an anxious disposition. If he were anxious, because of some internal mental conflict, he would not be steady, and he would shoot badly. We expect a man who is a "good shot" to finish off other jobs well. Of a number of men going in for a final examination, one will tremble, one will vomit, one will have diarrhoea, one will be restless, many will feel nicely excited, and a few will go in and sail through.

The anxious one who trembles is the one who might as a child have been sent to a doctor as ?-chorea; especially if the anxiety occurred even apart from examinations and externally-imposed ordeals, if it were liable to appear at any time of the day or night, the result of a mental conflict.

This is the child who shrinks from the spatula, so that you either have to give up the attempt to examine the throat or else use so much coercion that you scarcely feel justified in insisting on getting your own way. The more this child gets to like you, even to trust you, the less may it become possible for him or her to allow you to put in the spatula and depress the tongue.

Diagnosis is made yet harder in that these children, as a result of disturbed sleep and of the mental conflict behind this symptom, are tired in the morning and suffer from rheumatic pains indistinguishable from the pains of subacute rheumatism.

Here, then, is a child with the complexion of a rheumatic child, because of vasomotor instability, with a form of growing pains, with tiredness, and moreover with a universal anxious fidgetiness closely resembling the choreic movements of acute rheumatism. On examination of the heart, too, there is found a rapid thumping action, and even dilatation.

And yet it is suggested that it is possible to separate these cases off and to say they are not liable to heart disease. In other words, it is suggested that the disease is emotional in origin and that treatment, if required, would be by appropriate analysis of the emotional state. This is never true of true chorea, which is, I assume, a rheumatic encephalitis.

The obvious symptom in these cases is the presence of frequent and intense night terrors, and the condition is an anxiety hysteria and neurosis.

It has been my lot to watch a very large number of these children untreated for several years, and my experience leads me to the conclusion that the condition is not rheumatic.

[The condition seems to correspond with what Sutherland refers to when he writes of the "normal heart of a nervous subject." (See Garrod, Batten and Thursfield, G. A. Sutherland's section on "Disorders and Diseases of the Heart," 1929 edition.) For a clear exposition of the opposite view see Poynton's Lettsomian Lectures, *Lancet*, 1928, ii, page 586.]

These cases are often treated for long periods as chorea and subacute rheumatism, and the non-development in them of rheumatic heart disease is sometimes taken as a sign of effectual treatment. If my supposition is correct the freedom from

heart disease is part of the picture. It appears from what Dr. Poynton writes that he does not expect these cases to develop endocarditis, though he considers the tachycardia to be rheumatic.

My experience is that when chorea by chance occurs in a child of this type the prognosis is not good. The inability to obtain relative heart rest owing to the over-acting of the heart from fear increases the difficulty of treatment of the carditis which may develop.

The children of this group are most healthy when allowed to do everything that a healthy child can do. They limit their own activities, and when they are tired they rest themselves. And if they do not rest no heart damage is done.

They may suffer from other anxiety symptoms, pain in the epigastrium or "round the heart," fainting, recurring migraine-like headaches, recurring abdominal colic, vomiting, diarrhoea; but these symptoms should only make the physician bolder in his diagnosis of anxiety neurosis, and more determined not to restrict activity in the way that he would and should do if the case were one of subacute rheumatism or slight chorea.

It must be remembered that the excitable and nervous children, especially the former, may have habit spasms. Also some children who are excitable are nervous and some who are of the chronically anxious type are also excitable. But the three types of emotional disorder seem to be sufficiently clearly defined to be described apart.

Tics and habit spasms may occur in a child who has true chorea. Even then they should be given their proper valuation. Usually they were present before the onset of the chorea. But even if the chorea has determined the onset of the habit movements, such movements could not have been brought out had the child's emotional development been normal. The basis for the tic must have been already present. The chorea symptoms and signs, on the

other hand, develop without relation to the emotional state. They are as definitely organic as the more disastrous lesions of encephalitis and poliomyelitis.

Severe chronic excitement and anxiety states are common in children and chorea must and does occasionally occur in such children. My experience up to the present does not make me at all certain that the teaching is correct that chorea is more likely to attack over-nervous than normal children. The teaching that rheumatism is more liable to appear as chorea than as joint rheumatism in a nervous child may possibly be true. Chorea is known to occur more commonly in girls than in boys, and the chronic anxiety state is also met with rather more frequently in girls.

If it should be proved that nervousness means proneness to chorea, then a definite new line of preventive treatment would be opened up; for diseases of emotional origin, especially the anxiety states, with anxiety as a symptom, are now capable of radical treatment.

This relationship needs more careful analysis by observers who are able and willing to recognize disease with an emotional basis as well as disease due to infection and other physical conditions.

CONCLUSION.

(1) It is essential to have a clear picture both of true chorea and of the types of case which commonly resemble but which are not chorea for the diagnosis of the doubtful case. In fact, by this means the number of doubtful cases can be reduced to quite a small percentage.

(2) As a basis for further observation and discussion it is assumed that there is no relationship between the two sets of disorders. On the one hand there is true chorea and the liability to other rheumatic symptoms and signs and to heart disease. On the other hand there are certain common disorders of emotional development. These do occasionally resemble chorea, but even

then they carry no special liability to heart disease. They are not related to rheumatic fever or to its subacute form. It will be well at present to assume that they do not even predispose to rheumatism or to chorea.

SOME RECENT WORK ON ACUTE INFECTIOUS DISEASES.

AN ADDRESS DELIVERED BEFORE THE WINDSOR AND
DISTRICT MEDICAL SOCIETY, ON FEBRUARY 20, 1929.

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THE subject which I have chosen for this afternoon, "Some recent work on acute infectious diseases," naturally covers a very wide field, and in the time at my disposal I shall be able to discuss only a few aspects of the question, and these mainly from the clinical standpoint.

I propose therefore to deal with the following diseases which I have ranged in alphabetical order, not in the order of their importance, viz., chicken-pox, diphtheria, measles, scarlet fever and vaccinia.

CHICKEN-POX.

The causal connection between herpes zoster and varicella, which since the first communication of von Bokay, professor of children's diseases at Budapest in 1892, has formed the subject of an extensive literature, is still a matter of dispute. Is the virus of herpes zoster the same as that of varicella, as von Bokay, Le Feuvre (of Buluwayo) and Netter (of Paris) seem to think, or are zoster and varicella quite distinct diseases, as we had hitherto been accustomed to regard them?

Three varieties of the association have been described. In the first and much the

commonest, an attack of zoster in one member of a family or other community is followed within a period of three weeks—usually twelve to fourteen days, corresponding to the incubation period of varicella—by an attack of chicken-pox in another member of the community. A much rarer association of zoster and varicella is the occurrence of chicken-pox in one individual followed by the appearance of zoster in another with whom he has been in contact, of which only ten cases were collected by Netter in 1920 and twenty-one by his son in 1921. Intermediate in frequency is the group of cases of which I recently saw and published an example, consisting in the concurrence of herpes zoster and varicella in the same individual (*Brit. Journ. Child. Dis.*, 1926, xxiii, 270). In such cases the unicist school maintains that the concurrence of the two eruptions is due to dissemination of a virus which is at first localized to the posterior ganglia, whereas the dualists hold that the concurrence of the eruptions is a pure coincidence.

Until recently I was inclined to side with Comby, the principal representative of the dualists, who regard the association of the two diseases as a mere coincidence; but I must confess that my belief was somewhat rudely shaken by the following case. Some months ago I was asked to see a young woman who had had an attack of chicken-pox followed by facial paralysis, and on inquiry into the source of infection was told that her younger sister had had a few spots some weeks before. On examining the child I found the characteristic pigmented scars of chicken-pox on the trunk. I finally asked the father if he had had shingles lately, and much to my surprise and, I must confess, gratification, he at once replied in the affirmative, and showed me the typical pigmented scars of a cervical herpes zoster which had appeared about a fortnight before his little girl's eruption. The moral of this story is that one should always make an inquiry as to the occurrence of a previous