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THE NATURE OF THE RHEUMATIC CHILD

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The rheumatic state in childhood has remained a profound and complex problem despite the observation and research of the last 60 years. There is little in the clinical story of the disorder as related in the textbooks of to-day which would break new ground for Cheadle, Goodhart, and Still. The problems which they stated—the nature of chorea and its relation to rheumatism, the aetiology of the rheumatic state and its relation to diathesis—still refuse solution. These clinicians would certainly remark that in the last 30 years interest and emphasis have shifted from the nature and reactions of the host to the characters of the invader, or, in Osler's parallel, from the soil to the seed. These bacteriological researches have produced much knowledge but no finality. A recent summary of them states:

"According to one view rheumatic fever is caused by an unknown virus, an infection which is readily activated by haemolytic streptococci, but which may at times be activated under other circumstances. According to the second view the streptococci are themselves the cause of the rheumatic fever, the rheumatic symptoms being regarded as a peculiar type of host response conditioned by sensitization. It is not yet possible to decide between these views. It is a question whether the antibody response to the streptococcus infection which causes the rheumatic flare-up shows a significant difference from that encountered in individuals who show no such flare-up." (Holt and McIntosh, 1940.)

The comparative failure of these researches demonstrates the need for returning to the preoccupations of 50 years ago, to the study of the soil rather than of the seed, for it is here that the key to this "peculiar type of host response" must eventually be found.

The Mind-Body Relationship

There are other reasons for returning to the study of the disposition of the rheumatic child. There is no disorder in which the emotional and the physical components seem more inextricably mingled. There is no disorder in which the attempt to separate the psyche from the soma, the functional from the organic, is more baffling. It is a long step from the undischarged emotion of the choreic child to the adult dying from congestive heart failure, and of many of the steps in this unhappy pilgrimage we are ignorant. We shall not, however, advance our knowledge by refusing to recognize that this emotional tension exists in the child or by denying the possibility that such nervous instability can both initiate and condition physical processes. Psychiatry cannot discover the link between the emotional tension and the physical disorder; it can do no more than explore fully the emotional background. The other approach to this mind-body relationship is by way of neurophysiology, and it does not seem likely to be more fruitful than the psychiatric approach. Sherrington (1940) has discussed this problem in his Gifford Lectures, "Man on his Nature," and he writes:

"There is, for instance, the condition 'anxiety.' None is, I suppose, more far-reaching as a warper of the mind. But where does neurophysiology contribute anything to the knowledge of the

norm from which anxiety causes departure, and what has cerebral physiology to offer on the whole subject of 'anxiety'? The psychiatrist has perforce to go on his way seeking things more germane to what he needs. . . . The mental is not examinable as a form of energy. That in brief is the gap which parts psychiatry and physiology."

The introduction of the electro-encephalogram gave hope to some that the elucidation of this profound problem was imminent, and of this record it has been written (Gordon, 1939):

"It is interesting to note that these changes in Berger's rhythm may be brought about not only by traumatic stimuli or by stimuli such as the shining of a bright light into the eye affecting the occipital cortex, but also by purely intellectual processes such as the effort to solve a mathematical problem."

It is obvious that not much can be hoped in this direction from a machine which makes little distinction between a crack on the head and the intellectual processes of an Einstein. It gives us *quantity* when we ask for *quality*, or, as Sherrington has brilliantly expressed it:

"It has brought us to the brain as a telephone exchange. All the exchange consists of switches. What we really wanted of the brain was, it would seem, the subscribers using the exchange—the subscribers with their thoughts, their desires, their anticipations, their motives, their anxieties, their rejoicings."

To write and talk, then, of "psychosomatic disorders" as is so often done to-day is to assume a bridge whose existence we but dimly discern and of whose nature we are ignorant. Such illnesses are better named disorders of personality, of the soil, or of diathesis. Yet we must not refuse validity, for these same reasons, to the psychological assessment of the personality of sick people before, during, and after their development of the physical signs of disease; for such studies, rightly interpreted, insist on a fundamental unity of mind and body—an integrity which can only be denied by materialists or mystics who have lost their way. Too often, however, in adults such studies have no therapeutic value. The process is irreversible. The arthritic joints, the ulcerated duodenum or colon, dominate the patient's psyche and the physician's efforts; but in children the lesion is, for the most part, subordinate to the growth of personality. The tail does not as yet wag the dog. The conditioning process of continuous adjustment to the environment goes on inexorably; but where the conditioning is too difficult or too rapid and where the adjustment is faulty, the parents, the physician, and the teacher can intervene to anticipate disaster and prevent it. The rheumatic state in childhood is to some extent a preventable disaster (since it occurs unusually in the well-to-do classes), and therefore the first questions to ask when we are confronted by a child with subacute rheumatism, with chorea, and with acute arthritis are: "Why did this child become rheumatic, and how can the aetiological factors be controlled?" To answer these questions is the further purpose of this paper.

What is a "Functional" Disorder?

It is necessary first to consider one other problem in nomenclature—the definition of a "functional" disorder. However this word is defined, it is still interpreted as denoting

something of which the sufferer should be ashamed and something for which the physician may justly feel impatience or contempt. So that if I define chorea as a functional disorder, as I do, the immediate reaction of many people is: "Nonsense! How can the poor child help itself?" It is necessary at present to include under the description of "functional" disorders that must ultimately prove to be of widely different aetiology: the psychoneuroses; cerebral disorders, such as epilepsy, migraine, and chorea; the allergic disorders; the disorders of plain muscle—bronchial, oesophageal, gastric, colonic, rectal, ureteric, arterial; the disorders of cardiac rhythm, such as paroxysmal tachycardia; and many others as yet unclassifiable.

All this is so unsatisfactory that attempts are constantly made to provide a more exact nomenclature. Of recent years it has become usual to talk of physiogenic and psychogenic components of such disorders. This has been of little value because we are often unable to distinguish, as in the effects of cerebral trauma, what is physiogenic from what is psychogenic. Moreover, everyone agrees that physical disease may produce psychological disorder, and most of us accept as a fact that psychological states both indirectly and directly induce physical disease. A man is depressed, anxious, and fatigued, and develops a carbuncle, influenza, pneumonia, or herpes zoster; most of us are prepared to relate his psychological state to his ensuing physical disorder, just as we are prepared to believe that certain kinds of emotional stress may form the background for physical diseases, such as duodenal ulcer, ulcerative colitis, exophthalmic goitre, and rheumatoid arthritis. Changes in nomenclature cannot usefully run ahead of ascertained fact; for thus judgment is clouded and counsel confused. We must rest content with the adjectives "functional" and "organic" until the facts allow us to make further distinctions between them. If, then, I define chorea as a "functional" disease I mean that, although it has organic associations, it has a psychogenic component and no known structural pathology.

*The Clinical Significance of "Growing Pains"

"Growing pains" is the traditional name given to the pains in the limbs which occur commonly in children during school age. If the name was apposite it must have been generally inferred that such pains were both common and insignificant, that many children suffered from them and were afterwards none the worse. Such an estimate of their importance was strongly deplored by all physicians in early decades of this century. G. F. Still (1912) wrote:

"The practical importance of recognizing the significance of these slight pains in the limbs in children—'growing pains' as they are too often called—can hardly be overrated. They may be the earliest indication of a rheumatic taint, and as such should always serve as a danger signal. To disregard them is to favour the onset of more severe symptoms, and it seems likely that active treatment in this early stage may not only cause the disappearance of pains but may also prevent those affections of the heart that are so distressing a feature in the rheumatism of childhood; certainly these vague pains call for the utmost care in prophylaxis, and a child who has once shown such symptoms requires continual care as to clothing, climate, and the prevention of exposure to cold and damp."

Robert Hutchison (1940) spoke more epigrammatically of "growing pains" in words which have echoed in the ears of several generations of medical students: "That, gentlemen, is a phrase which has crippled a great many hearts."

This was the general view, taught, accepted, and practised until after the last war, when the L.C.C. started its clinics for rheumatic children at the children's hospitals in London. Great numbers of children with pains in the limbs were referred to these clinics as suffering from incipient or actual rheumatic disease. It was soon obvious to the physicians in charge that they were not suffering from the rheumatic state; nor did they, in any significant number, develop it while under observation (Winnicott, 1931; Sheldon, 1936; Hawksley, 1939). Attention, both here and in America, during the last 10 years has therefore turned to the nature of these pains, their frequency, and their differentiation from the rheumatic state. They are variously regarded by different authorities as being due to 'normal physiologic growth'

* The part of this paper concerned with growing pains was read to the Derby Medical Society on August 11, 1942.

(Shapiro, 1939), a view widely held in the nineteenth century; to fatigue and damp (Sheldon, 1936); to fibrositis (Seham and Hilbert, 1933); to postural deformities (several authors); and to emotional disorders (Winnicott, 1931, 1938). No one has yet attributed them to a deficiency of the vitamin B complex. Two of these views may be considered before discussing the nature of these pains. That the pains may be due to normal physiological growth is an opinion which will commend itself to few. The only physiological process to be accompanied by pain is childbirth, and to regard pain as a proper concomitant of growth is to live in a crazy world where the normal and abnormal coincide.

Nor is it to be accepted that fatigue is the prime cause of these pains. It is true that tiredness is frequently complained of by these children, but this fatigue is itself a symptom of ill-health. There is no fatigue in a healthy child that is not recoverable from by a night's rest. Tired children are ill children, and their illness may be emotional or physical in origin.

The incidence of these growing pains in normal children is extraordinarily high. Some observers have suggested that it is as high as 20% and even 40%; but a careful and extensive inquiry made by Kaiser (1927) in New York showed that out of 48,000 normal school children 7.1% complained of pains in the limbs. The incidence of cardiac rheumatism among 301,392 L.C.C. children examined in 1936 was 0.8%; so we may conclude that the incidence of growing pains among children is nine times greater than the incidence of rheumatic heart disease.

If the problem of the relation of growing pains to rheumatism is considered from another angle an immediate difficulty is disclosed, for there is no doubt that children who have developed rheumatic heart disease often give a history of pains in the limbs. Kaiser (1934) in his account of an investigation of 1,200 children with rheumatism said that 18% complained of muscle pains; and Sheldon reported that nearly 20% of 266 children with rheumatic heart disease gave a history of aches and pains in the extremities.

This, then, is the justification that the older physicians would have advanced for their warning as to the profound danger of growing pains. Their remedy was to treat all children with growing pains as potential rheumatics if their hearts were normal and as actual rheumatics if their hearts were under suspicion. This meant, as the quotation above from Still demonstrates, a life of partial invalidism, and it is not surprising, therefore, that the incidence of cardiac neurosis in the adult population is greater than the incidence of rheumatic heart disease. Paul Wood (1941) has written graphically of the childhood of these cardiac neurotics:

"On the approach of the evil day when they will have to attend school they are filled with apprehension; if the dire event is not delayed by some happy illness (rheumatic fever followed by an open-air school is especially welcomed) they spend the next few years as trembling spectators of the rough-and-tumble of the miniature world about them. They evade as much as possible with the help of friendly coughs and colds; kindly doctors and soft mothers are at hand to protect them from the hazards of football, swimming, and the gymnasium."

As there is nothing in the disorders of school children that has more importance than this attitude of doctors and parents, let me recount the story of a pilgrim who narrowly escaped from this slough of despond:

Case 1.—This patient was 17 years old when I first saw him. His condition had been diagnosed as subacute rheumatism at the age of 12 by a well-known paediatrician on account of tachycardia and pains in the limbs. After this he became completely invalid. He was not allowed to go to school, but had a tutor at home. He was forbidden all games and swimming. He was kept in bed for a fortnight whenever he caught a cold, which he frequently did. His parents surrounded him with every circumstance of anxiety—they were convinced that his heart was "weak." Fortunately, it was not so difficult to convince the boy that his heart was normal. He started at 17 to swim and play tennis, and without his parents' knowledge he joined the Territorials just before the war. The start of the war found him in the ranks, and the violent change in his conditions of life was almost too much for him. He fainted several times on parade. Fortunately, the boy had spirit and the makings of a good physique, but his parents during the next year tried every manœuvre to get him out of the Army.

His father's heart bled for him metaphorically, but his duodenal ulcer bled literally. Fortunately again, my opinion of his heart was firmly supported by Dr. Hugh Barber and Dr. Geoffrey Bourne. The boy remained in the Army, and some time ago successfully completed his Commando training.

The moral of this story is not that the original diagnosis was incorrect. It may possibly have been right. The moral is that such treatment is hopelessly bad even for a child with rheumatic heart disease.

Pitfalls in Diagnosis

Can this diagnosis between the child with active rheumatism and the child with growing pains be made with reasonable certainty? No doubt it can, but the following characteristic examples will illustrate the pitfalls in diagnosis and the errors in treatment.

Case 2.—A boy aged 10 complained of pains in his legs. His parents had kept him in bed as he was running a temperature between 99° and 101°. There was no evidence of infection, his heart was healthy, and his joints showed no inflammation. He had a mild secondary anaemia. He said that his pains were in his calf and thigh muscles. He slept badly, was difficult to control, cried frequently, and attacked his young sister without good reason. We kept him in bed some weeks without improvement, and then at his parents' instance took him to see a well-known paediatrician. He spent ten minutes looking at the boy, declared him healthy and slapped him on the back, told the parents to send him away to boarding school, and ignored me completely. The boy returned to health and remained well.

I learnt from this salutary experience that emotional disorder in children can be established as a positive diagnosis and not a diagnosis made simply by exclusion of physical signs. The only diagnostic sign in this boy was his emotional instability. The second lesson was that emotional disorder can cause signs and symptoms that are often thought only to be physically determined—pyrexia, tachycardia, secondary anaemia, and pains.

Case 3.—A boy aged 13 was referred from school clinic on account of a pain in his left elbow which had been present for a year. He was a healthy and alert boy of good physique with a sensible and cheerful mother. His pain frequently came on at school, and it was so severe that he often wept and had to be sent home. Equally, it might come on at home, as "last night when he was writing a letter to his father." He was very much attached to his father, who joined the Army 14 months ago. Pain frequently waked him at night and he slept badly. Six months ago he was seen by an orthopaedic surgeon, who found no disease but referred him for physiotherapy. There was no improvement after four months' treatment. No sign of ill-health was elicited on examination, and he was treated by *reassurance* that his pain betokened no disease; *explanation* that the pain was due to depression at his father's absence; *insistence* that he should carry on with what he was doing at home or at school, however great the pain; *sedative* (phenobarbitone gr. 1/2 night and morning) to ensure sleep and a quiet nervous system. These simple measures brought about relief from pain in a fortnight, and the boy has remained well.

This boy illustrated one point that is commonly overlooked: the necessity for removing the presenting symptom—i.e., pain—in these children. Quite often it seems that the emotional disorder which originally produced the symptom has disappeared and the child's entire preoccupation is with the pain. Remove the pain or give the child sleep and all is well.

Case 4.—A girl aged 8 was referred from the school medical department with pains in the legs and ? early chorea. The child was evacuated from Birmingham and now billeted with a foster-mother, who brought her to hospital. Her movements were continuous as the woman told the story of her fidgetiness, irritability, and sleeplessness. The movements were not characteristic of chorea. Her temperature was 99.5°, her pulse rate 120 a minute. The apex beat was in the nipple line, but there was a short systolic murmur at the apex. She had a kypho-lordotic stance, and her foster-mother, facing the child while I looked at her back, went on with her garrulous repetition of the patient's defects. The child's spine suddenly twitched into a well-marked scoliosis and then went back as suddenly to the midline again. She was admitted for a few days for observation away from her foster-mother. Her symptoms settled quickly; no further organic signs were discerned; her sedimentation rate and her sleeping pulse rate were normal. The billet-

ing officer found another home for her, and she remained well after discharge.

These last two cases raise the question of orthopaedic treatment. It is wrong, as in Case 1, to expect the orthopaedic surgeon to diagnose emotional disorder. One must not hope to "gather figs from thistles," but the orthopaedist is not without sin in treating by physiotherapy a pain which showed no orthopaedic associations. The last child, who was beginning postural deformities, might have needed orthopaedic treatment provided the emotional difficulty was coincidentally treated. The emotional origin of some postural deformities is increasingly recognized by orthopaedic surgeons. Bankart has written: "To find a scoliosis look at the child's back; to find its cause look at its mother's face." Hawksley (1939) found that "a frequent cause of growing pains is a postural or orthopaedic defect such as flat-foot, knock-knee, scoliosis, or bad stance; treatment of the cause in these cases nearly always gave relief."

I agree that it is fairly common to find a postural deformity in these children, but I do not agree that this is commonly a source of their growing pains. I have often caused the disappearance of growing pains in these children (as in Case 4) without treating their postural deformity. I agree again that physical exercise is good treatment for such children: the improvement of physical poise brings a coincident improvement in mental poise.

Growing Pains and the Subacute Rheumatic State

In Case 4, above, the subacute rheumatic state was very closely imitated. The pains, the temperature, the tachycardia, and the systolic murmur suggested a rheumatic carditis, and the fidgetiness and habit spasms mimicked true chorea. Such cases are not very uncommon, and it is admitted by all clinicians that an absolute diagnosis may be difficult. In this case a positive diagnosis of emotional disorder, the absence of evidence of an active carditis, and the therapeutic test of removal from environmental difficulties provided the correct solution. Yet it is such cases that the older physicians had in mind when they wrote of a "pre-rheumatic state," of "a latent rheumatic infection or toxæmia," of "subacute rheumatism," or of "latent chorea": a state which had for its presenting symptoms muscle or joint pains, fatigue, pallor, and an emotional instability.

This idea of latent infection or toxæmia makes little appeal to a generation which is inclined to discount the kindred theory of the septic focus. An infection which cannot be demonstrated either clinically or bacteriologically cannot be assumed to be present. Moreover, the notion that such cases show evidence of a pre-rheumatic state is open to two overwhelming objections—viz., (1) that all nervous children show these and similar symptoms; (2) that to treat such children as rheumatic is to make cardiac neurotics of them.

"But," the older physicians would reply, "a definite percentage of children with an active carditis associated with rheumatism give a history of what we have called subacute rheumatism." This is true, but in some of these children the symptoms are probably due to a coincident and active carditis. The number of children in whom the first and presenting symptom of the rheumatic state was growing pains alone *without* carditis was 9% in Findlay's series of 701 rheumatic children and 8% in Kaiser's series of 1,200. Shapiro (1939) suggests that it is possible by analysis of the type of pain felt by these children to make a diagnosis between subacute rheumatism and growing pains. It is obviously of the highest importance to attempt this diagnosis in the individual child, since growing pains do not lead to carditis and the pains of subacute rheumatism may do so. Shapiro concludes that growing pains come on at the end of the day or at night, ~~waking~~ waking the child, that they are situated in the muscles of the thighs, and that the children show none of the other evidences of rheumatic activity; while the pains of subacute rheumatism are present in the morning and persist throughout the day, are made worse by walking and are situated in the joints themselves, and such children show other evidence of rheumatic activity. The most valuable sign in this differential diagnosis is the situation of the pain. If the pain is limited to joints, even in the absence of swelling, subacute rheumatism

should be suspected. All agree that in a small number of cases immediate diagnosis is impossible, and Shapiro writes:

"In a smaller group of patients who complain of painful extremities a clear-cut differentiation cannot be made. They have symptoms which suggest both the muscular pain of 'growing pains' and specific joint pain as well, together with some signs of infection. Such children need to be observed over a period of time before a definite diagnosis can be made."

More helpful in the positive diagnosis of growing pains is the presence of emotional disorder. The child is depressed or anxious, and shows this not alone by growing pains but also by behaviour disorders and disturbance of body function—sulkiness, irritability, fits of temper, sleeplessness (almost invariably present), fatigue, and lack of appetite. The therapeutic test of simple psychotherapy and sedation often confirms the diagnosis. There is now general agreement by physicians who have watched large numbers of these children with growing pains over many years that they do not, in any significant number, develop rheumatism. Sheldon (1936) examined 189 children with growing pains after a lapse of four years and found that only two had developed carditis. On the one hand, then, there is the fact that 9% of children with active carditis give a history of subacute pains which it may sometimes be difficult to distinguish from growing pains, and on the other the fact that of vast numbers of children under observation for growing pains only a negligible proportion develop active rheumatism. Can these two facts be reconciled?

I think it is wise to keep a more open mind on this question than is generally done to-day. It is generally agreed that children with growing pains who are under proper observation and treatment do not develop rheumatism. Is it certain that the same can be said of those children whose pains are neglected and in whom emotional disorder persists? I have notes of three children whom I had previously diagnosed as suffering from growing pains and who then disappeared from my sight. They reappeared in the out-patient department, after attacks of acute tonsillitis, at intervals varying from six months to two years, each of them with an active carditis. The fact that there is sometimes, as Shapiro and all other observers agree, this difficulty in deciding between the two conditions makes one wonder still whether this state of subacute rheumatism may not be an intermediate phase, in children predisposed to rheumatism, between growing pains and active rheumatism.

This matter of subacute rheumatism awaits clarification; it should be emphasized that it is an uncommon way for the child to enter into the rheumatic state. In so far as I have seen subacute rheumatism it has been easier to detect in those children evidence of nervous instability than to find unequivocal evidence of infection. In the effort to diagnose active rheumatism at the earliest opportunity there is no doubt that these cases have been given an importance out of proportion to their comparative infrequency. They have a great aetiological interest, however, for they raise the questions—Is there sometimes a half-way house to active rheumatism?; Could the process at this stage be controlled?

The Nature of Growing Pains

Four of the theories which have been advanced for the causation of growing pains—normal growth, fatigue, latent infection, and postural deformities—have been discarded already in the course of this paper. That growing pains may be due to fibrositis finds few supporters: Hawksley discovered only one or two cases among his 115. Tenderness at the site of the pain is remarkably absent in these children. Damp as a contributory factor can also be excluded in most cases. The remaining theory is Winnicott's suggestion that these pains represent an emotional disorder associated with the child's growth. "They are to be found," he writes, "wherever human beings are developing from babies into children, from children into adolescents, and from adolescents into adults." I have accepted Winnicott's theory as a basis for treatment in the many scores of such children I have seen in the last six years, and I am firmly convinced of its truth. These children's pains respond to the simplest psychotherapeutic and sedative measures, to the great improvement of their physical and nervous health. Statistics bring no conviction in this regard—it is a matter for personal approach and experience; but

statistical evidence is provided by Neustatter (1937), who examined from the psychiatric standpoint a group of 40 children with growing pains, against several groups of controls. He reported that 34 out of 40 children with limb pains showed some degree of nervous disorder, as against 15 out of 40 picked children of good physique and 22 out of 40 in a group of rheumatic children. He concluded that these children with growing pains were "suffering from the effects of prolonged emotional strain."

It is common to find evidence in support of this theory of "prolonged emotional strain" in the papers of authors who attribute growing pains to other causes. Many observers have noted the fact that these children cry frequently, unlike the children with acute rheumatism. Hawksley says: "45% of the 115 patients with growing pains had at one time or another been reduced to crying with the pain. A feature noted by many parents was the tendency of growing pains to make their children sulky and irritable. Psychological maladjustment was unexpectedly frequent, resulting in unhappy school or home life."

It is concluded, then, that growing pains in children reflect a disorder of psychological adjustment and they are quickly removed by simple and appropriate treatment. Active rheumatism should never be diagnosed unless arthritis or carditis is present. The possibility that subacute rheumatism represents the growing pains of children predisposed to rheumatism cannot be denied. This consideration is only of theoretical importance in aetiology, and is of no account in the treatment of growing pains.

The Nature of Chorea

Chorea is generally defined as a manifestation of the rheumatic state whose morbid anatomy is an inflammation of some part of the cerebrum. It is open to anyone to draw a conclusion from the known facts, but the usual inference made from this definition is that chorea is a brain inflammation caused directly by the rheumatic invader. This definition and the inferences from it differ so remarkably from a legitimate conclusion based on the available evidence that a reconsideration of the facts is overdue. In the first place, chorea is a disorder whose structural pathology is quite unknown.

"Microscopical changes," write Buzzard and Greenfield (1921), "in the central nervous system in cases of chorea have usually been conspicuous by their absence, but attention has been called to the occasional presence of hyperaemia and of small thrombotic lesions which, however, are by no means constant."

Lewy—quoted by Findlay (1931): "Summing up the position in 1924, Lewy states quite categorically that changes are very seldom found, that these are of no definite nature and are no more than are to be observed in the brains of patients dying from any acute disease."

Freeman (1933) makes no distinction between the morbid anatomy of the brain in acute rheumatism and chorea. He classes the changes in the brain with those seen in typhus and the rickettsias.

Holt and McIntosh (1940) write: "There has been little constancy in the findings in patients who have died with chorea, and there is still some doubt as to the seat of the morbid process. Degenerative changes in the basal ganglia, the cortex, and the cerebellum have been described by Lewy and others in a number of instances. In other cases no such lesions have been found although carefully searched for, and one must assume that the disturbance is functional."

These authors define chorea as "a functional disorder of the nervous system characterized by sudden aimless, irregular movements often accompanied by muscular weakness and emotional instability." Kinnier Wilson (1928) writes: "Choreic motor disorder manifests itself through the pyramidal tracts, which must be in a condition of relative integrity for hyperkinetic symptoms to appear, and yet they exhibit some defect of function."

Chorea is a disorder which to-day steadily diminishes in frequency and severity. Even in days when it was more common and more violent Still records that "there are exceedingly rare cases in which it proves fatal by its violence. . . . Almost invariably if death occurs during chorea it is due to endocarditis or pericarditis." A survey of recently published necropsies suggests that the morbid anatomy described depends on the supervening rheumatic inflammation. The changes recorded in the brain are widespread from pons to cortex, and it can only be said of such changes that they are what would be expected of acute rheumatism by comparison

with the morbid anatomy of other organs, and if these extensive lesions of meningo-encephalitis are proffered for the structural basis of uncomplicated chorea then they bear little relation to the clinical picture of the disorder.

That chorea is a functional nervous disorder unassociated with brain inflammation is borne out by clinical observation. It is accompanied by no pyrexia, by no leucocytosis, by no signs of inflammation in the cerebrospinal fluid, and by no raising of the sedimentation rate unless carditis is present. The movements of chorea cease completely during sleep, unlike the similar movements that are induced by encephalitis. Recovery from chorea is complete and there are never any organic nervous sequelae. The uncomplicated case of chorea runs its course, from onset to recovery, without one clinical or laboratory sign which indicates an infection or inflammation. I know of no other disease in which, without some positive evidence, inflammation and infection are generally assumed to be present.

It is possible that invasion of the brain by a virus, activated on the one hand by emotional stress and on the other by the haemolytic streptococcus, may give rise to the emotional and motor instability which is called chorea, without revealing its penetration by the usual signs of infection. This is an adequate and attractive theory, but without proof. It is accepted by most physicians that chorea is often preceded by a prolonged emotional strain, such as the "anxieties engendered in nervous children by the examination system" or by domestic stresses or, more rarely, by a sudden shock or fright. It occurred to Findlay that the "chorea induced by fright might be different in nature from that arising spontaneously; but of the 36 cases with a history of fright cardiac disease was present in 47%, and of the 254 cases in which no history of fright was obtained cardiac disease was also present in 47%, so that there seems little justification for such an assumption." It would be possible to quote from a dozen authors who deny that such emotional disorders have any aetiological significance in the development of chorea: believing as they do that chorea is caused by a rheumatic infection, these physicians cannot be expected to agree that it may also be initiated by the school inspector or by domestic infelicity. They therefore suggest that an attack by a dog, or whatever it may be, is too trivial an incident to occasion such a disorder. This will be generally accepted. The fright has the same relation to the ensuing chorea as the murder of the Archduke at Sarajevo had to the development of war in 1914. This view that chorea was often emotionally induced was firmly held by nineteenth-century physicians, and it is accepted as firmly by most physicians to-day.

Sheldon (1936) writes: "The important part played in the production of chorea by the stress of education cannot be doubted. There is often a history of some shock or fright at the beginning of an attack."

Carey Coombs (1924) states: "Nothing is more certain than that nervous stress precedes the onset of chorea in a number of cases—a number too large to be explained away."

Winnicott (1938) says: "In my own clinic I had no difficulty in collecting 50 cases of chorea following acute or prolonged emotional strain."

Gerstley *et al.* (1935), after a close clinical, psychiatric, and social study of 58 children in Chicago suffering from chorea, writes: "Psychic trauma seemed so intimately related to the actual onset in 22 cases as to force the question of its being the activating factor. Domestic discord was a prominent feature. It was plainly apparent in 32 families and extreme in 13. The onset of acute manifestations often followed a domestic crisis such as divorce, separation, desertion, drunkenness, and the advent of stepbrothers and step-sisters. Other types of emotional injury were common. In three girls the onset was related to attempted rape. In four others the child lived in continuous dread of assault by undesirable characters in the neighbourhood. One child was aware of the illicit sexual relations of her mother. In others the onset followed acute fright. In short, in our series psychic trauma resulting from fright or overwhelming grief stood out far more strikingly than did any history of infection."

A reflex of the Chicago underworld in the early 1930's! Of the 18 cases of chorea seen by me during the last five years four started acutely after a sudden fright. One child developed chorea actually during an air raid, in one it followed a few hours after a sexual assault, in one after an attack by a dog, and in the fourth after the child had been called to the front of the class by a school inspector and asked to recite. In six of the other children there was evidence of

prolonged emotional stress either at home or at school. One child developed it after tonsillitis, and in three other children its onset accompanied an attack of acute rheumatism.

Two other facts in the aetiology of chorea concern social grouping and sex incidence. The social grouping in chorea is even more marked than in acute rheumatism. Most physicians can report instances of acute rheumatism among children of the well-to-do classes (see below), but chorea is rarely present among these children. Still (1927) saw only one case of chorea in his consulting practice in 700 consecutive cases between the ages of 6 and 10 years. Chorea is three times as common in girls as in boys. Still (1912) attributes this to the greater nervous instability of girls, and here few will be found to disagree. This remarkable disparity in sex incidence is not shown by rheumatic arthritis or carditis.

The Relation of Chorea to Rheumatism

The facts concerning the relationship of chorea to rheumatism may be concisely stated. They are that (1) rather more than one-quarter of all rheumatic children enter the rheumatic state by way of chorea; (2) in addition, another quarter of all rheumatic children develop chorea at some time in the evolution of their rheumatism; and (3) a number of children with chorea, estimated by various authorities to be between one-third and one-half, never develop any rheumatic episode.

These facts mean that chorea is a frequent associate of the rheumatic state in children. In some children the choreic state never progresses to rheumatism; in others the chorea precedes the rheumatic state by weeks, months, or years; while in others chorea develops during the phase of acute rheumatism or at some time after it. The inference from these facts is that the effect of the rheumatic inflammation on the cerebrum of a predisposed child is of exactly the same nature as the effect of prolonged emotional strain or acute psychic trauma. It is suggested that all these factors operate by heightening an emotional and kinetic instability which is already present in the child. It is apparent that the association of chorea with rheumatism is without parallel in clinical medicine. The profound difficulties in understanding this association come near to solution if the rheumatic state is regarded as a unique disorder with two components—nervous instability on the one hand and an inflammation of the heart and joints on the other. Both of these components may occur independently, but either, in some unexplained way, favours the development of the other. In any case the facts concerning chorea do not support any theory that chorea is directly due to the rheumatic "invader" or to "rheumatic toxæmia." It may be regarded either as a prelude to the rheumatic state or its accompaniment; but frequently, and fortunately, it is neither the one nor the other.

Chorea, then, may be defined as a functional disorder of the nervous system, characterized by irregular motor activity and emotional instability, which develops in predisposed individuals, particularly girls of the poorer classes, following acute psychic trauma or prolonged psychic stress, or during and after the development of the rheumatic state.

(To be concluded. A list of references will appear at the end of Part II)

The Minister of Health announces (Circular 2734) that he has had under review the arrangements for the supply of insulin free or at a reduced price to persons suffering from diabetes. Under the National Health Insurance Acts an insured person entitled to medical benefit may obtain insulin free if it is prescribed or dispensed by his insurance doctor. Insulin may also be provided as part of medical relief for any destitute person under the Poor Law Acts. As regards school children, in a few cases the Board of Education has in the past sanctioned the supply of insulin free or at a reduced price where the parents could not afford the whole cost. This sanction is now extended so that all education authorities may, for the duration of the war, supply insulin free or at a reduced price to all children at elementary schools or at institutions for higher education. The needs of other persons for whom there is no public provision apart from poor relief, such as dependants of insured persons, widows and spinsters engaged in household duties, and merchant seamen on foreign-going ships, are to be met by a wider use of the powers given to local authorities under the Public Health Act, 1936, temporarily to supply medicine to the poorer inhabitants of the district. The Minister now gives the necessary consent to the supply of insulin to such persons for the duration of the war.