patient was in a hurry. This is sometimes in striking contrast to the scanning speech of insular sclerosis. The fingers are flexed and in the position assumed when the hand is at rest; in the late stages they cannot be extended. Occasionally there is overextension of the terminal phalanges. The hand is usually turned toward the ulnar side, and the attitude somewhat resembles that of advanced cases of rheumatoid arthritis. In the late stages there are contractures at the elbows, knees, and ankles. The movements of the patient are characterized by great deliberation. He rises from the chair slowly in the stooping attitude, with the head projecting forward. In attempting to walk the steps are short and hurried, and, as Trousseau remarks, he appears to be running after his centre of gravity. This is termed festination or propulsion, in contradistinction to a peculiar gait observed when the patient is pulled backward, when he makes a number of steps and would fall over if not prevented—retropulsion.

The reflexes are normal in most cases, but in a few they are exaggerated.

Of sensory disturbances Charcot has noted abnormal alterations in the temperature sense. The patient may complain of subjective sensations of heat, either general or local—a phenomenon which may be present on one side only and associated with an actual increase of the surface temperature, as much as 6° F. (Gowers). In other instances, patients complain of cold. Localized sweating may be present. The mental condition rarely shows any change.

Variations in the Symptoms.—The tremor may be absent, but the rigidity, weakness, and attitude are sufficient to make the diagnosis. The disease may be hemiplegic in character, involving only one side or even one limb. Usually these are but stages of the disease.

Diagnosis.—In well-developed cases the disease is recognized at a glance. The attitude, gait, stiffness, and mask-like expression are points of as much importance as the oscillations, and usually serve to separate the cases from senile and other forms of tremor. Disseminated sclerosis develops earlier, and is characterized by the nystagmus, and the scanning speech, and does not present the *attitude* so constant in paralysis agitans. The hemiplegic form might be confounded with post-hemiplegic tremor, but the history, the mode of onset, and the greatly increased reflexes would be sufficient to distinguish the two. The Parkinsonian face is of great importance in the diagnosis of the obscure and anomalous forms.

The disease is incurable. Periods of improvement may occur, but the tendency is for the affection to proceed progressively downward. It is a slow, degenerative process and the cases last for years.

Treatment.—There is no method which can be recommended as satisfactory in any respect. Arsenic, opium, and hyoscyamia may be tried, but the friends of the patient should be told frankly that the disease is incurable, and that nothing can be done except to attend to the physical comforts of the patient.

Other Forms of Tremor.

(*a*) *Simple Tremor.*—This is occasionally found in persons in whom it is impossible to assign any cause. It may be transient or persist for an indefinite time. It is often extremely slight, and is aggravated by all causes which lower the vitality.

(*b*) *Hereditary Tremor.*—C. L. Dana has reported remarkable cases of hereditary tremor. It occurred in all the members of one family, and beginning in infancy it continued without producing any serious changes.

(*c*) *Senile Tremor.*—With advancing age tremulousness during muscular movements is extremely common, but is rarely seen under seventy. It is always a fine tremor, which begins in the hands and often extends to the muscles of the neck, causing slight movement of the head.

(*d*) *Toxic tremor* is seen chiefly as an effect of tobacco, alcohol, lead, or mercury; more rarely in arsenical or opium poisoning. In elderly men who smoke much it may be entirely due to the tobacco. One of the commonest forms of this is the alcoholic tremor, which occurs only on movement and has considerable range. Lead tremor will be considered in speaking of lead poisoning, of which it constitutes a very important symptom.

(*e*) *Hysterical tremor*, which usually occurs under circumstances which make the diagnosis easy, will be considered in the section on hysteria.

III. ACUTE CHOREA

(*Sydenham's Chorea; St. Vitus's Dance*).

Definition.—A disease chiefly affecting children, characterized by irregular, involuntary contraction of the muscles, a variable amount of psychical disturbance, and a remarkable liability to acute endocarditis.

We shall speak here only of Sydenham's chorea. Senile chorea, chronic chorea, the prehemiplegic and post-hemiplegic forms, and rhythmic chorea are totally different affections.

Etiology.—*Sex.*—Of 554 cases which I have analyzed from the Philadelphia Infirmary for Diseases of the Nervous System, seventy-one per cent were in females and twenty-nine per cent in males. After puberty the percentage in females increases.

Age.—The age incidence in 522 cases was as follows: In the first decade, 201; in the second decade, 248; in the third decade, 10; in the fourth decade, 1; above the fourth decade, 2. In the cases under twenty years the following is the age incidence in the hemidecades: In the first hemidecade, 33; in the second hemidecade, 168; in the third hemidecade, 212; in the fourth hemidecade, 52.

Station.—While the disease affects children of all grades of society, it is more common among the lower classes.

in their efforts by teachers and parents, form a large contingent of the cases of chorea in hospital and private practice. Sturges has called special attention to this *school-made* chorea as one serious evil in our modern method of forced education. *Imitation*, which is mentioned as an exciting cause, is extremely rare, and does not appear to have influenced the onset in a single case in the Infirmary records.

The disease may rapidly follow an injury or a slight surgical operation. Reflex irritation was believed to play an important *rôle* in the disease, particularly the presence of worms or genital irritation; but I have met with no instance in which the disease could be attributed to either of these causes. Local spasm, particularly of the face—the habit chorea of Mitchell—may be associated with irritation in the nostrils and adenoid growths in the vault of the pharynx, as pointed out by Jacobi.

It has been claimed by Stevens that *ocular defects* lie at the basis of many cases of chorea, and that with the correction of these the irregular movements disappear. To test the truth of these statements a careful study was made at the Infirmary by De Schweinitz of the condition of the eyes in 50 cases of chorea in children, with the following results: Hypermetropia was present in 23, or forty-six per cent; hypermetropia in one eye and hypermetropic astigmatism in the other in 7, or fourteen per cent; hypermetropic astigmatism in 12, or twenty-four per cent; myopia in 1, or two per cent; myopic astigmatism in 3, or six per cent; mixed astigmatism in 4, or eight per cent. De Schweinitz then adds the cases reported by Stevens and C. S. Bull, of New York, making a total of 227 cases, of which 112 were ametropic and 115 emmetropic. His conclusions are as follows: "Hypermetropia and hypermetropic astigmatism are vastly the preponderating condition in the eyes of choreic children, being found in about seventy-seven per cent of the cases, exactly as hypermetropic refraction is the preponderating condition in childhood, being found in seventy-six per cent of the eyes of children in the elementary schools," and the "evidence, however, seems quite as lacking that hypermetropic refraction is the basal cause of chorea, as it is that the chorea is the cause of the hypermetropia."

The committee of the New York Neurological Society which investigated with great care and impartiality Stevens's claims came to the conclusion that the facts did not warrant their adoption.

Morbid Anatomy and Pathology.—No constant lesions have been found in the nervous system in acute chorea. Vascular changes, such as hyaline transformation, exudation of leucocytes, minute hæmorrhages, and thrombosis of the smaller arteries, have been described.

Embolism of the smaller cerebral vessels has often been found, as might be expected in a disease with which endocarditis is so frequently associated. Based upon this fact, Kirkes, Tuckwell, Hughlings Jackson, and Bastian have supported what is known as the embolic theory of the disease. Endocarditis is by far the most frequent lesion in Sydenham's

chorea. With no disease, not excepting rheumatism, is it so constantly associated. In the records of over 110 autopsies, in nearly 100 this condition was mentioned. In the 5 autopsies of which I have notes, in all the mitral valves were affected. The endocarditis is usually of the simple variety, but the ulcerative form has occasionally been described.

We are still far from a solution of all the problems connected with chorea. Unfortunately, the word has been used to cover a series of totally diverse disorders of movement, so that there are still excellent observers who hold that chorea is only a symptom, and is not to be regarded as an etiological unit. The chorea of childhood, the disease which Sydenham described, presents, however, characteristics so unmistakable that it must be regarded as a definite, substantive affection. We cannot discuss fully, but only indicate briefly, certain of the theories which have been advanced with regard to it. The most generally accepted view is that it is a functional brain disorder affecting the nerve-centres controlling the motor apparatus, an instability of the nerve-cells, brought about, one supposes by hyperæmia, another by anæmia, a third by psychical influences, a fourth by irritation, centric or peripheric. Of the actual nature of this derangement we know nothing, nor, indeed, whether the changes are primary and the result of a faulty action of the cortical cells or whether the impulses are secondarily disturbed in their course down the motor path. The predominance of the disease in females, and its onset at a time when the education of the brain is rapidly developing, are etiological facts which Sturges has urged in favor of the view that chorea is an expression of functional instability of the nerve-centres.

The embolic theory originally advanced by Kirkes and supported by the English writers above mentioned has a solid basis of fact, but it is not comprehensive enough, as all of the cases cannot be brought within its limits. There are instances without endocarditis and without, so far as can be ascertained, plugging of cerebral vessels; and there are also cases with extensive endocarditis in which the histological examination of the brain, so far as embolism is concerned, was negative. In two of my post-mortems there were certainly no emboli in the smaller arteries of the branches of the circle of Willis or of the cortex. In the third there was a spot in one corpus striatum of red softening, probably due to an embolus. In favor of the embolic view is the experimental production in animals of chorea by Rosenthal, and later by Money, by injecting fine particles into the carotids of animals.

Lately, as indeed might be expected, a microbic origin has been sought for, and, however improbable such a theory looks at first sight, the case of tetanus gives a warrant, at least, to speculation and investigation in this direction. Nothing definite has yet been determined. From Naunyn's clinic a case is reported with endocarditis and a reddish-brown infiltration of the pia at the base of the brain which proved to be a microbic growth similar in character to those in the vegetations on the heart

valves. Recently, in a fatal case in my wards cultures of a micrococcus were obtained from the blood of the heart, and throughout the brain there were minute foci of hæmorrhage similar to those which occur in pneumonia and other infectious disease associated with endocarditis. In favor of this view it has been urged, as it is impossible to refer the chorea to endocarditis or the endocarditis in all cases to rheumatism, that both have their origin in a common cause, some infectious agent, which is capable also, in persons predisposed, of exciting articular disease. Cases have been reported in scarlet fever with arthritic manifestations, in puerperal fever, and rheumatism, also after gonorrhœa, and such facts are suggestive at least of the association of the disease with infective processes. Possibly, as has been suggested by some writers, the paralytic conditions associated with chorea may be analogous to those which occur in typhoid and certain of the infectious diseases. On the other hand, there are conditions extremely difficult to harmonize with this view. The prominent psychical element is certainly one of the most serious objections, since there can be no doubt that ordinary chorea may rapidly follow a fright or a sudden emotion. It cannot be supposed, too, that the forms associated with reflex irritation, as from the nose and particularly the cases of so-called habit chorea, can be dependent upon infection. We must place these in a separate category, and yet in a long series cases shade so imperceptibly into each other that it is extremely difficult to separate them properly. The question deserves careful study, and the possibility of a special infectious agent has of late been advocated by several writers.

Symptoms.—Three groups of cases may be recognized—the mild, severe, and maniacal chorea.

Mild Chorea.—In this the affection of the muscles is slight, the speech is not seriously disturbed, and the general health not impaired. Premonitory symptoms are shown in restlessness and inability to sit still, a condition well characterized by the term "fidgets." There are emotional disturbances, such as crying spells, or sometimes night-terrors. There may be pains in the limbs and headache. Digestive disturbances and anæmia may be present. A change in the temperament is frequently noticed, and a docile, quiet child may become cross and irritable. After these symptoms have persisted for a week or more the characteristic involuntary movements begin, and are often first noticed at the table, when the child spills a tumbler of water or upsets a plate. There may be only awkwardness or slight incoördination of voluntary movements, or constant irregular clonic spasms. The jerky, irregular character of the movements differentiates them from almost every other disorder of motion. In the mild cases only one hand, or the hand and face, are affected, and it may not spread to the other side.

In the second grade, the *severe form*, the movements become general and the patient may be unable to get about or to feed or undress herself, owing to the constant, irregular, clonic contractions of the various muscle

groups. The speech is also affected, and for days the child may not be able to talk. Often with the onset of the severer symptoms there is loss of power on one side or in the limb most affected.

The third and most extreme form, the so-called maniacal chorea, or *chorea insaniens*, is truly a terrible disease, and may develop out of the ordinary form. A young girl, aged eighteen, was admitted to the Montreal Hospital October 17. She was a waitress at a hotel, and being badly frightened by two men who were fighting, she dropped a tray of dishes which she was carrying. A severe reprimand increased her worry and trouble. The next day she packed her trunk and went home, a distance of thirty miles. Her father insisted that she should return. At this time her hands and arms began to twitch in a violent manner. Five days after the first fright she was admitted. The arms and legs were in constant motion, jerking in all directions. The face also was affected. She was rational, but could scarcely speak. On the night of the 19th she had no sleep, but raved and talked all the time, and the movements were incessant. On the 20th, 21st, and 22d the condition persisted and grew worse. The temperature ranged from 101° to 103°, the tongue became dry and cracked, and she became much exhausted. On the night of the 22d the temperature rose to 105° and death followed, ten days after the onset of the symptoms. These cases are more common in adult women and may develop during pregnancy.

Chorea begins, as a rule, in the hands and arms, then involves the face, and subsequently the legs. The movements may be confined to one side —hemichorea. The attack begins oftenest on the right side, though occasionally it is general from the outset. One arm and the opposite leg may be involved. In nearly one fourth of the cases speech is affected; when slight this is only an embarrassment or hesitancy, but in other instances it becomes an incoherent jumble. In very severe cases the child will make no attempt to speak. The inability is in articulation rather than in phonation. The lips and tongue are concerned in the defect. Occasionally the inspiratory muscles are involved, even when the speech is not at all affected, and sobbing and sighing may result. Paroxysms of panting and of hard expiration may occur, or odd sounds may be produced. As a rule the movements cease during sleep.

A prominent symptom is muscular weakness, usually no more than a condition of paresis. The loss of power is slight, but the weakness may be shown by an enfeebled grip or by a dragging of the leg or limping. In his original account Sydenham refers to the "unsteady movements of one of the legs, which the patient drags." There may be extreme paresis with but few movements—the paralytic chorea of Todd. Occasionally a local paralysis or weakness remains after the attack. Case 229 of the Infirmary series, a lad of ten, had severe general chorea in September, 1880, with considerable loss of power in the legs. Recovery was slow, and when he returned in September, 1883, in a second attack of chorea, there was talipes of the

left foot, which had resulted from paralysis in 1880. In Case 21 a wrist-drop persisted for two years, the result of a palsy which came on with chorea. These are probably instances of peripheral neuritis.

A question of some interest is whether choreic spasms extend to the muscles of organic life. The great gastro-intestinal muscle is never affected. There are no symptoms which can be referred to anomalous contractions of the stomach or bowels. The sphincters act normally. Incontinence of urine occurs occasionally, but it is not noted more frequently, I think, in chorea than in other nervous affections. Spasm of the bronchial muscles is not found even in severe cases, in which the respiratory muscles are involved. The pupils are usually dilated, but no irregular contractions occur. The rapid action and disturbed rhythm of the heart present nothing peculiar to the disease, and there is no support for the view that irregular contractions occur in the papillary muscles.

Heart Symptoms.—*Neurotic.*—As so many of the subjects of chorea are nervous girls, it is not surprising that a common symptom is rapidly acting heart. Any emotional disturbance causes at once a marked increase in the number of the beats, and the actions may become irregular and tumultuous. Irregularity, however, is not so special a feature in chorea as rapidity. The patients seldom complain of pain about the heart.

Hæmic Murmurs.—With anæmia and debility, not uncommon associates of chorea in the third and fourth week, we find a corresponding cardiac condition. The impulse is diffuse, perhaps wavy in thin children. The carotids throb visibly, and in the recumbent posture there may be pulsation in the cervical veins. On auscultation a systolic murmur is heard at the base, perhaps, too, at the apex, soft and blowing in quality.

Endocarditis.—As in rheumatism, so in chorea, acute valvulitis rarely gives evidence of its presence by symptoms. It must be sought, and clinical experience has shown that it is usually associated with murmurs at one or other of the cardiac orifices.

For the guidance of the practitioner the following statements may be made:

(1) In thin, nervous children a systolic murmur of soft quality is extremely common at the base, particularly at the second left costal cartilage, and is probably of no moment.

(2) A systolic murmur of maximum intensity at the apex, and heard also along the left sternal margin, is not uncommon in anæmic, enfeebled states, and does not necessarily indicate either endocarditis or insufficiency.

(3) A murmur of maximum intensity at apex, with rough quality, and transmitted to axilla or angle of scapula, indicates an organic lesion of the mitral valve, and is usually associated with signs of enlargement of the heart.

(4) When in doubt it is much safer to trust to the evidence of eye and hand than to that of the ear. If the apex beat is in the normal position, and the area of dulness not increased vertically or to the right of the sternum, there is probably no serious valvular disease.

(5) The endocarditis of chorea is almost invariably of the simple or warty form, and in itself is not dangerous; but it is apt to lead to those sclerotic changes in the valve which produce incompetency. Of 110 choreic patients* examined more than two years after the attack, 54 presented signs of organic heart-disease.

(6) Pericarditis is an occasional complication of chorea, usually in cases with well-marked rheumatism.

Sensory Disturbances.—Pain in the affected limbs is not common. Occasionally there is soreness on pressure. There are cases, usually of hemichorea, in which pain in the limbs is a marked symptom. Weir Mitchell has spoken of these as *painful choreas*. The pain may be quite apart from any arthritic complications. Tingling and pricking sensations and numbness are found occasionally. Anæsthesia is very uncommon. Tender points along the lines of emergence of the spinal nerves or along the course of the nerves of the limbs are rare. The French writers have compared these to the hysterogenic points in hysteria, and have also described in certain cases ovarian tenderness. Headache may be a very troublesome symptom.

Psychical disturbances are common, though in a majority of the cases slight in degree. Irritability of temper, marked wilfulness, and emotional outbreaks may indicate a complete change in the character of the child. There is deficiency in the powers of concentration, the memory is enfeebled, and the aptitude for study is lost. Rarely there is progressive impairment of the intellect with termination in actual dementia. Acute melancholia has been described (Edes). Hallucinations of sight and hearing may occur. Patients may behave in an odd and strange manner and do all sorts of meaningless acts. By far the most serious manifestation of this character is the maniacal delirium, occasionally associated with the very severe cases—*chorea insaniens*. Usually the motor disturbance in these cases is aggravated, but it has been overlooked and patients have been sent to an asylum.

The psychical element in chorea is apt to be neglected by the practitioner. It is always a good plan to tell the parents that it is not the muscles alone of the child which are affected, but that the general irritability and change of disposition, so often found, really form part of the disease.

The condition of the *reflexes* in chorea is usually normal. Sinkler made observations at the Philadelphia Infirmary in 50 cases with the following results: In 26 the knee-jerk was normal, in 15 it was diminished in degree, and in 9 it could not be obtained. Trophic lesions rarely occur

* American Journal of the Medical Sciences, 1887, ii.