

## **On chorea and choreiform affections / by William Osler.**

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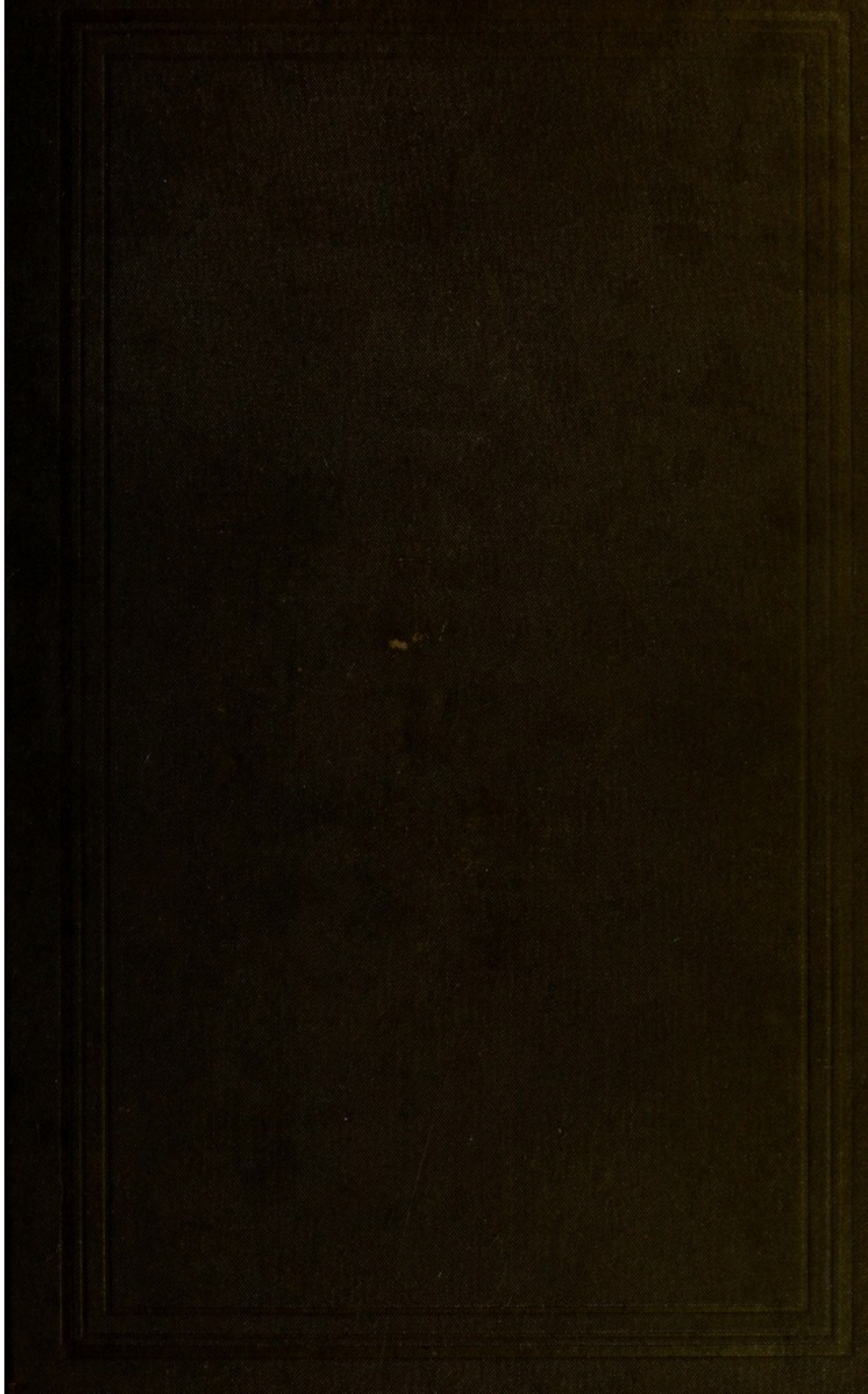
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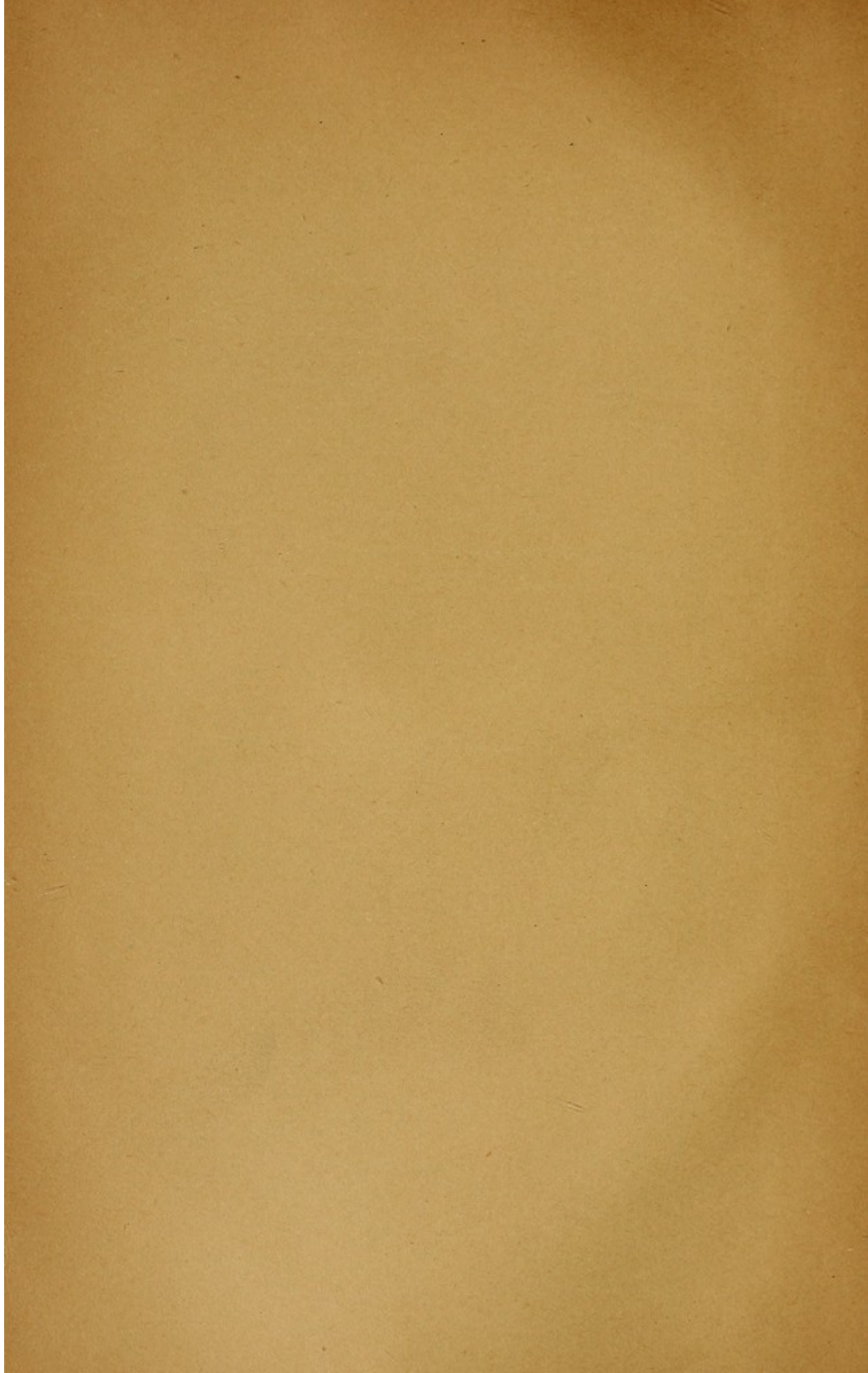
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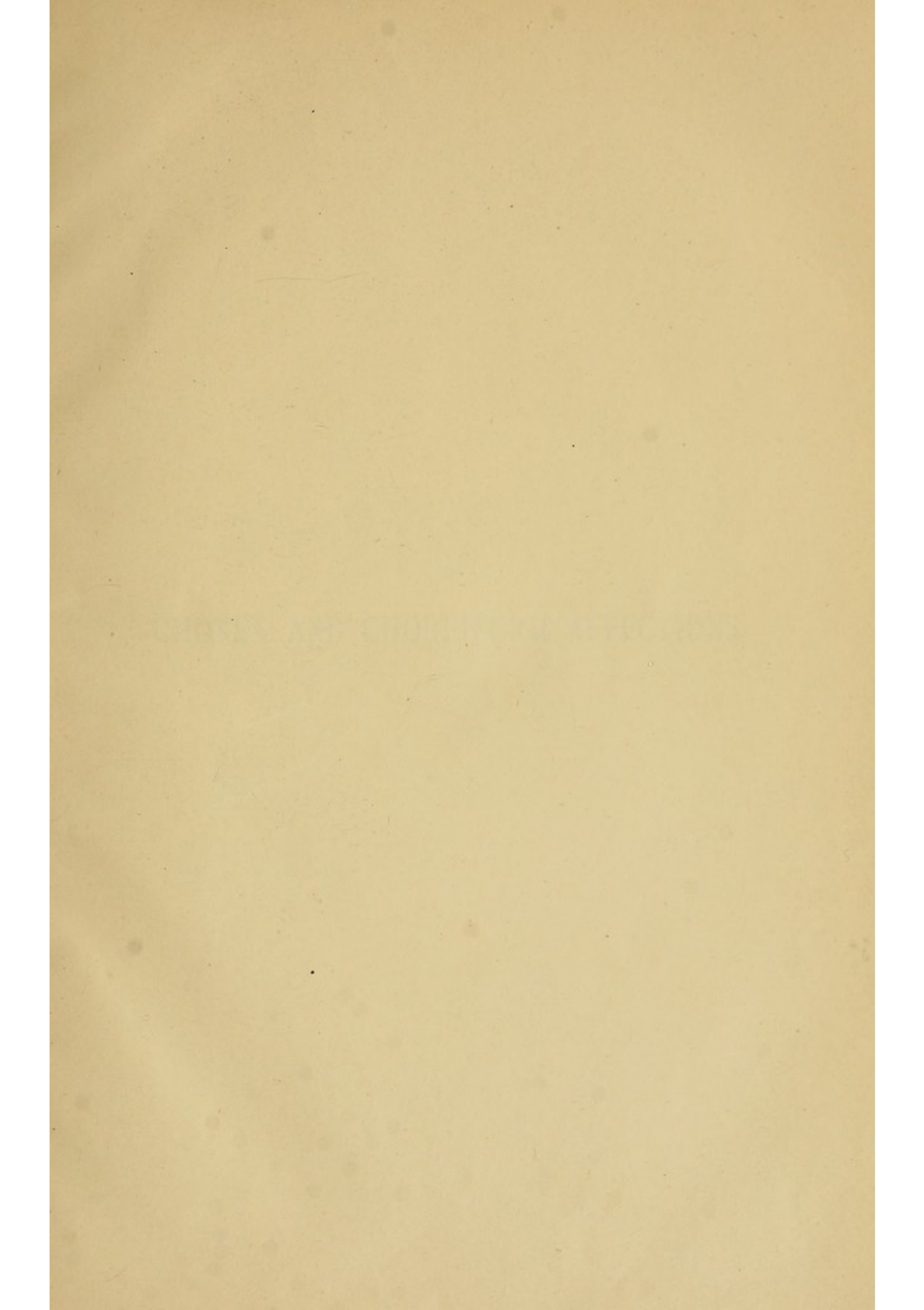




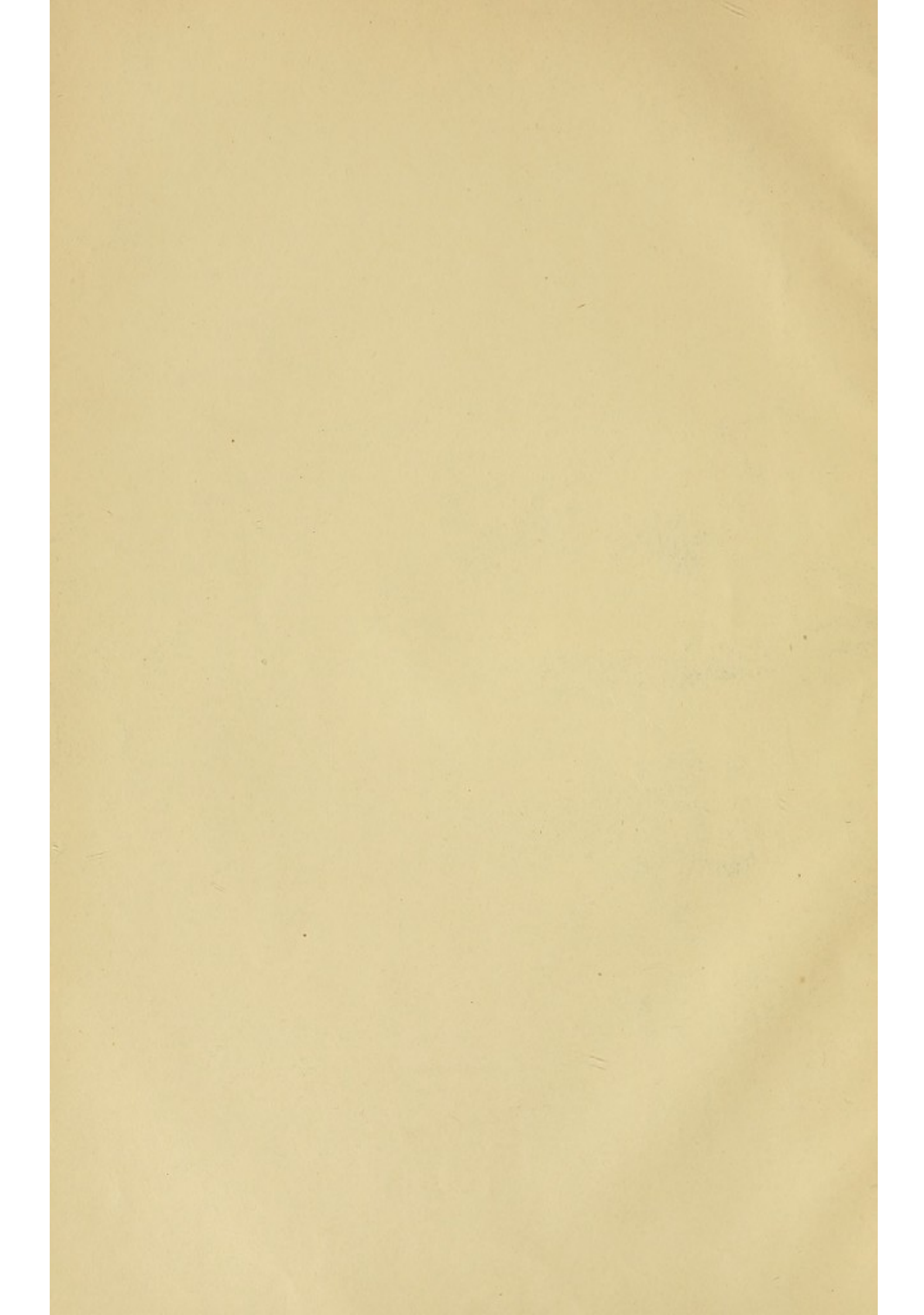






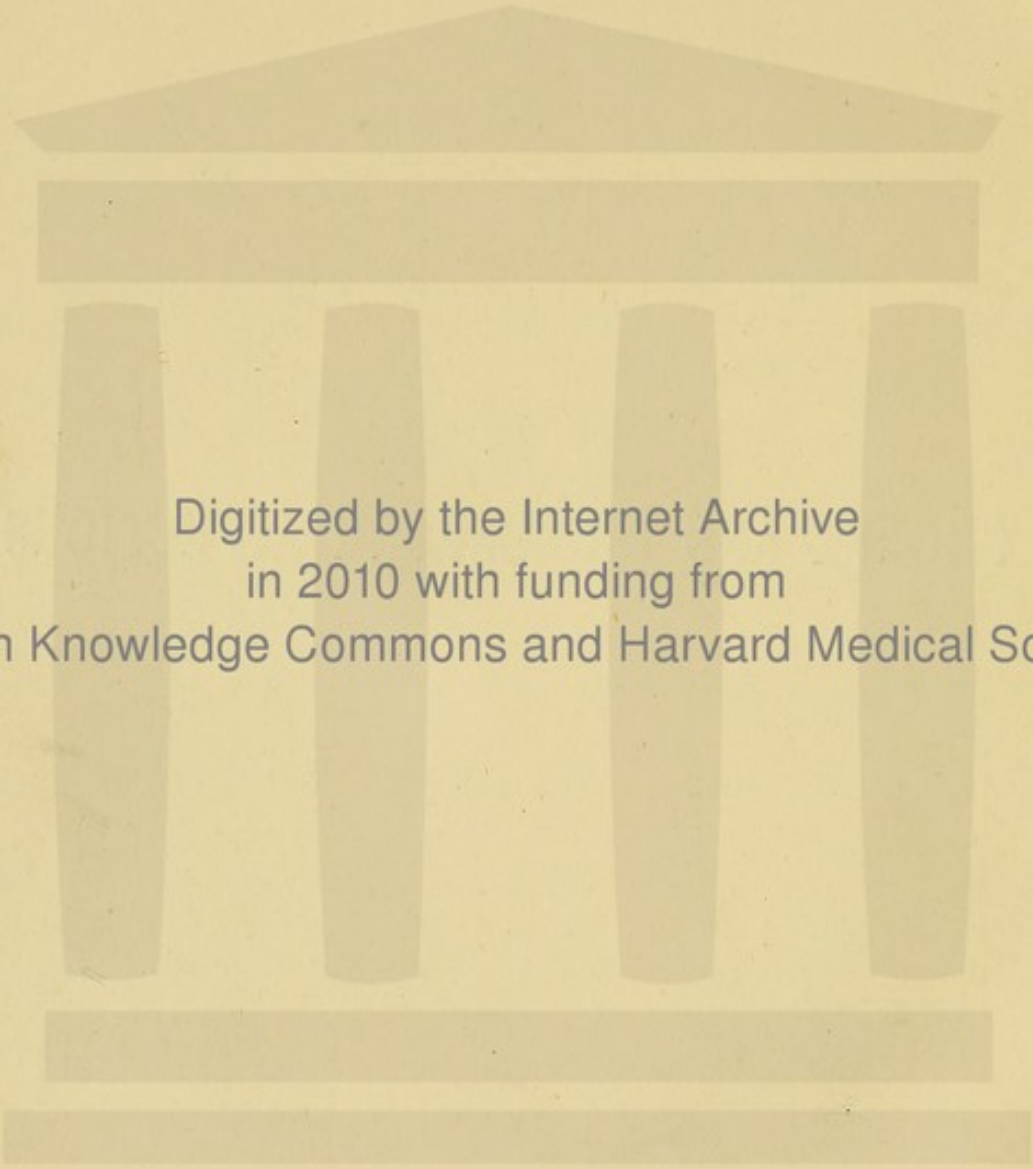






ON  
CHOREA AND CHOREIFORM AFFECTIONS





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ON

CHOREA

AND

CHOREIFORM AFFECTIONS

BY

WILLIAM OSLER, M.D.

FELLOW OF THE ROYAL COLLEGE OF PHYSICIANS, LONDON; PRESIDENT OF THE ASSOCIATION  
OF AMERICAN PHYSICIANS; PROFESSOR OF MEDICINE JOHNS HOPKINS UNIVERSITY,  
AND PHYSICIAN-IN-CHIEF JOHNS HOPKINS HOSPITAL, BALTIMORE.  
FORMERLY PHYSICIAN TO THE INFIRMARY FOR DISEASES OF THE NERVOUS SYSTEM,  
PHILADELPHIA

PHILADELPHIA

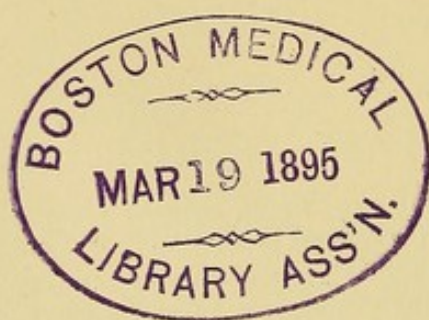
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1894



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TO W. R. GOWERS, M.D., F.R.S.

DEAR GOWERS,

To the profession of the United States and Canada you stand as the most brilliant British exponent of the complex science of neurology.

Please accept the dedication of this little volume as an earnest of the gratitude felt towards you by thousands of your kinsmen across the water, and as an expression of the personal attachment of

Your sincere friend,

THE AUTHOR.

"Tout est extraordinaire dans cette Maladie; son nom est ridicule, ses symptômes singuliers, son caractère équivoque, sa cause inconnue, son traitement problématique."

—BOUTEILLE, in Preface of his "Traité de la Chorée," Paris, 1810.

"Vix datur morbus ullus qui toties medicorum de sua natura et indole illuserit judica ut Chorea Sti. Viti."

—BERNT, "Monographia Choreæ Sti. Viti," Prague, 1810.

## P R E F A C E

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A VERY large part of the material utilized in these studies is from the case-books of the Infirmary for Diseases of the Nervous System, Philadelphia. The sections on the general etiology and symptomatology of Chorea minor represent, in expanded form, the Lectures on Chorea which appeared in the *Medical News*, 1887. The chapter on *The Heart in Chorea* is also based on the Infirmary cases and on a careful study of a series of autopsies collected from the literature since 1881.

I have to express my obligations to my former colleagues, Dr. Weir Mitchell, Dr. Wharton Sinkler, and Dr. Morris J. Lewis, who permitted me to use their cases in preparing the statistical details of the 554 cases of chorea minor which are in the books to May 1st, 1889; also to Dr. Charles Burr and to Dr. Caspar Sharples for great assistance in the tabulation of the cases, and more particularly for their aid in the study of the condition of the heart in old patients.

JOHNS HOPKINS UNIVERSITY,

*July 1st, 1894.*





# CONTENTS

---

	PAGE
DEDICATION . . . . .	v
PREFACE . . . . .	vii

---

INTRODUCTION . . . . .	i
------------------------	---

## CHAPTER I.

Historical Note—Etiology, Age, Sex, Race, Seasonal Relations, Imitation, Trauma, Reflex Irritation, Eye-strain—Rheumatism (Arthritis)—Infec- tious Diseases—Hysteria—Poisons . . . . .	3
--	---

## CHAPTER II.

### SYMPTOMS.

Mild, Severe, and Maniacal Forms—Chorea Insaniens—Special Symptoms —Motor Disturbances—Muscular Weakness . . . . .	20
---	----

## CHAPTER III.

### SYMPTOMS (*continued*).

Disturbances of Sensation—Headache—Mental Symptoms—Cutaneous Symptoms—Urine, Nephritis—Fever—Duration—Recurrence—Chorea in Pregnancy—Chorea Minor in the Aged . . . . .	33
---	----

## CHAPTER IV.

### THE HEART IN CHOREA MINOR.

Condition of the Heart during the Attack—Condition of the Heart in Fatal Cases—Subsequent Heart History in Choreic Patients—Pericarditis in Chorea . . . . .	47
--	----

## CHAPTER V.

## MORBID ANATOMY AND PATHOLOGY—TREATMENT.

	PAGE
Statistics of Fatal Cases—Changes in the Nervous System—Bacteriology— Pathology—Treatment . . . . .	60

## CHAPTER VI.

## CHOREIFORM AFFECTIONS—HABIT SPASMS—TIC.

i. Simple Tic—Habit Spasm or Habit Chorea—Generalized Tic. ii. Tic with Imperative Ideas, &c. iii. Complex co-ordinated Forms of Tic. iv. Cases of Noisy Spasm of Respiratory Muscles . . . . .	72
---	----

## CHAPTER VII.

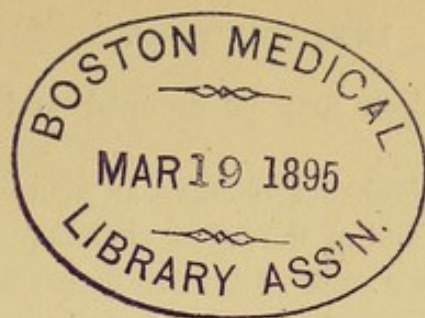
## CHRONIC PROGRESSIVE CHOREA (HUNTINGTON'S CHOREA).

Historical Note—Report of Two Families—Special Symptoms—Morbid Anatomy—Diagnosis. . . . .	96
--	----

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APPENDIX : TABLE OF RECENT AUTOPSIES . . . . .	113
--	-----





ON

# CHOREA

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## INTRODUCTION

IN the whole range of medical terminology there is no such *olla podrida* as Chorea, which for a century has served as a sort of nosological pot into which authors have cast indiscriminately affections characterized by irregular, purposeless movements. With muscular disorder as the salient feature, as the generic character, of chorea, there have been scores of specific designations, indicating the quality of the movement, the locality involved, &c. In Foster's Dictionary ninety-four of these sub-varieties are named. In the gradual growth of our knowledge of spasmodic affections this confusion has been perhaps inevitable. Even to-day it is not possible to make a satisfactory etiological classification, and the best we can do is to separate certain well-defined clinical forms, to which we may attempt to limit the use of the term chorea.

The epidemic disorder of motion which Paracelsus called Chorea Sancti Viti has a sort of prescriptive right to the name. Sydenham's error in adopting it for an affection of a totally different nature has been condoned by two centuries of usage; so that to these two forms, known respectively as chorea major and chorea minor, and each as St. Vitus's Dance, the name will doubtless cling. Then comes a long series of motor disorders in which the term has been freely used—the habit spasms and the various forms of tic so often confounded with chorea minor, the so-called symptomatic choreas, the chronic, the hereditary, the congenital, and the spastic forms, and the pre- and post-hemiplegic disorders of motion.

These various affections may be grouped and defined as follows :—

**Chorea Minor**—Sydenham's chorea—an acute disease of childhood, rarely of adults and of the aged, characterized by irregular, involuntary movements, a variable amount of psychical disturbance, and associated very often with arthritis and endocarditis. The disease is usually regarded as a neurosis, but the clinical characters of the severer cases, and the frequent heart and joint implication, have suggested to many recent writers that it may be due to a specific poison.

**Chorea Major**, under which term are now embraced both the dancing mania and the various forms of rhythmical or hysterical disorders of motion. Psychical impressions, emotional disturbances, and imitation play the most important *rôle* in this form.

**Choreiform Affections** or **Pseudo-Choreas**.—The various forms of habit spasm or tic, local or generalized, which are perhaps best grouped under the latter term, in the more extended use as employed by the French.

**Secondary** or **Symptomatic Choreas**.—Chronic disorders of motion, which depend upon degenerative and irritative lesions of the motor cortex or path. Here may be included the pre- and the post-hemiplegic disorders of movement, the so-called spastic choreas, and many of the cases of congenital and chronic chorea. One malady alone in this group may be separated as an independent affection, viz., the chronic progressive form, the so-called Huntington's chorea.

I shall not consider here chorea major, which belongs to hysteria, or the varieties known as symptomatic, except the chronic progressive form.



## CHAPTER I.

## HISTORICAL NOTE. ETIOLOGY.

Historical Note—Etiology, age, sex, race, seasonal relations, imitation, trauma, reflex irritation, eye-strain—Rheumatism (arthritis)—Infectious diseases—Hysteria—Poisons.

**Historical Note.**—The recognition of chorea minor as a distinct disease dates from Sydenham, whose *Schedula Monitoria* (1686) and *Processus Integri* (1693) contain brief but very accurate accounts of the affection as we now know it. His description in the *Schedula Monitoria* is as follows:—"St. Vitus's dance is a sort of convulsion which attacks boys and girls from the tenth year until they have done growing. At first it shows itself by a halting, or rather an unsteady movement of one of the legs, which the patient *drags*. Then it is seen in the hand of the same side. The patient cannot keep it a moment in its place, whether he lay it upon his breast or any other part of his body. Do what he may it will be jerked elsewhere convulsively. If any vessel filled with drink be put into his hand, before it reaches his mouth he will exhibit a thousand gesticulations like a mountebank. He holds the cup out straight, as if to move it to his mouth, but has his hand carried elsewhere by sudden jerks. Then, perhaps, he contrives to bring it to his mouth. If so, he will drink the liquid off at a gulp; just as if he were trying to amuse the spectators by his antics.

"Now this affection arises from some humour falling on the nerves, and such irritation causes the spasm."<sup>1</sup>

During the fourteenth, fifteenth and sixteenth centuries, under the influence of religious excitement, there were epidemics of dancing mania in Germany and the Netherlands. Pilgrimages were made to various shrines in search of relief, and as that of St. Vitus, in Zabern, was especially famous the dance became known by his name. Elsewhere, from other shrines, the disease received other names,

<sup>1</sup> *Sydenham's Works*. Latham's translation. Vol. II, p. 198. Sydenham Society's Edition.



as St. John's, or St. Anthony's dance. The epidemics gradually died out, but, curiously enough, as Charcot states, an annual pilgrimage of dancers, in perpetuation of the custom, is still made to the church of St. Willibrod, possibly the same as the "procession of the jumping saints" which Hirsch says is still commemorated at Echternach in Luxemburg. It is interesting to note that in Kentucky and Tennessee, in the early part of the century, during periods of intense religious enthusiasm, there were epidemic convulsions, similar in some respects to the dancing mania of the middle ages. In 1805, Dr. Felix Robertson described them in an "Essay on Chorea Sancti Viti," in his doctorate thesis at the University of Pennsylvania.

For a full and accurate description of the dancing mania, as it has occurred in various countries, Hecker's work<sup>1</sup> is still the standard authority. He there states that Paracelsus, in the beginning of the sixteenth century, made the disorder a subject of special study, and gave to it the name chorea. Though he made three divisions, they all referred to the dancing mania or to the hysterical form. It was unfortunate, to say the least, that Sydenham should have given the name chorea to an affection which had nothing whatever to do with the Chorea Sancti Viti, but custom has now sanctioned the use; and this is not the only instance in medicine in which we know a disease by a name the original significance of which has long been lost.

The modern study of the disease dates from the monographs of Bouteille<sup>2</sup> in France, and of Bernt<sup>3</sup> in Bohemia. The work of Bouteille is in many respects remarkable. In addition to a very full and accurate historical study, he gives an admirable clinical description of the disease, and makes a useful classification into essential or proto-pathic, secondary or deuterio-pathic, and false or pseudo-pathic. The cases are given in great detail, and he appears to have been fully aware of the association in many instances with rheumatism.<sup>4</sup>

<sup>1</sup> *The Epidemics of the Middle Ages*. J. F. C. Hecker, M.D., Sydenham Society, London, 1844.

<sup>2</sup> *Traité de la Chorée*, par E. M. Bouteille. Paris, 1810.

<sup>3</sup> *Monographia Choroæ Sti. Viti*, auctore Josepho Bernt, Med.D., &c. Praga, MDCCCX.

<sup>4</sup> The work is in another respect remarkable as the production of an octogenarian. In the dedication of the work to Vicq-D'Azyr he speaks of himself as *octogenarius medicus*. He was born in 1732, graduated from Montpellier, and was Government Physician for the study of miliary fever, on which he wrote a memoir. He practised at Mansosque in the department of the Basses-Alpes. He wrote on many subjects, particularly on hydrophobia. He died in 1815, in his eighty-fourth year.



Bernt's monograph is of special value, as it contains the older literature, and the reports of many interesting cases. All sorts of convulsive disorders are described under the name chorea. Many of the names still current, of varieties and sub-divisions of the disorder, of which he gives thirty-one ! date from this work.<sup>1</sup>

In France the brilliant lectures of Trousseau, but especially the monograph of Sée in 1850, and the extensive article of Roger, were among the most important contributions. Charcot and his pupils have contributed enormously to the proper appreciation of the varieties of chorea, and to our knowledge of the choreiform affections.

In Germany Romberg, Steiner, von Ziemssen, and many others, have published important studies.

In another than the usual sense, the disease may be called Chorea Anglorum, since by far the most important contributions have been made by english physicians. From Guy's Hospital have come many notable contributions. Bright, in Volume II. of the *Reports on Medical Cases*, 1831, gives an admirable clinical description of the disease, and gives a special section on chorea and rheumatism. He also called attention to the association of pericarditis with chorea. In the *Guy's Hospital Reports* are papers by Babbington, Vol. VI., 1841 ; by H. M. Hughes, 1846 ; by H. M. Hughes and E. Burton Brown, 1855 ; by Lever (on Chorea of Pregnancy), 1847 and 1848 ; by Pye-Smith, 1874 ; and by Goodall, 1890 ; and other contributions have been made by Wilks and Hilton Fagge. Of other British writers who have contributed extensively to our clinical and anatomical knowledge of the disease, may be mentioned Todd, Begbie, Kirkes, Ogle, Hughlings-Jackson, Broadbent, Dickinson, Tuckwell, Bastian, Sturges, Duckworth, Gee, Gowers, Money, Stephen Mackenzie, and Herringham. In the United States there have been reports of cases and careful studies by Levick, H. C. Wood, Mills, Jacobi, Hamilton, Haven, Putnam, and others ; while from the Philadelphia Infirmary for Diseases of the Nervous System, a large number of important contributions have been made by Weir Mitchell, Sinkler, Gerhard, Allison, Morris J. Lewis, and De Schweinitz.

<sup>1</sup> Bernt was born in 1770, graduated from Prague in 1797, was made Professor of Forensic Medicine in Prague in 1808. In 1813 he was called to Vienna to the same chair. He was the author of many works on Medical Jurisprudence.



### GENERAL ETIOLOGY.

The records of the Philadelphia Infirmary for Diseases of the Nervous System to May 1st, 1889, contain 554 cases, upon which the statements here made are based, supplemented to some extent by reference to cases at the Dispensary for Diseases of the Nervous System, Johns Hopkins Hospital.

**Sex.**—Females are attacked in the proportion of rather more than two to one; thus of 554 cases 161 were males, and 390, about 70 per cent., were females. In three the sex was not noted. This is a lower proportion than is given by many authors. After puberty the preponderance of females is much greater than in children.

**Age.**—It is essentially a disease of childhood and adolescence. The age incidence of 522 cases is given in the following table:—

	Totals.	Under 4	5	6	7	8	9	10	11	12	13	14	15	16-20	21-25	26-30	31-35	36-40	Over 40
MALES .	137	7	3	10	11	9	20	11	21	2	14	5	11	11	1	...	...	...	1
FEMALES .	385	11	12	15	28	42	43	39	37	29	19	23	25	51	5	4	1	...	1
	522	18	15	25	39	51	63	50	58	31	33	37	36	62	6	4	1	...	2

Arranged in decades the figures are as follows:—

First decade . . . . .	261
Second decade . . . . .	248
Third decade . . . . .	10
Fourth decade . . . . .	1
Above fourth decade . . . . .	2

Arranged in hemi-decades the figures are:—

First hemi-decade . . . . .	33
Second hemi-decade . . . . .	228
Third hemi-decade . . . . .	212
Fourth hemi-decade . . . . .	62

It is thus seen that the second and third hemi-decades are the periods in which occur more than three-fourths of the entire number of cases. In females the age incidence is somewhat earlier than in males. The second hemi-decade contains the greatest number of cases in males, and the third the greatest number in females. The



age incidence seems to be somewhat earlier in the United States than in Great Britain. In the report of the Collective Investigation Committee of the British Medical Association,<sup>1</sup> by Stephen Mackenzie, of the 439 cases analyzed, the largest proportion occurred in the third hemi-decade. Practically the disease is rare under the fourth year. The Infirmary records include two cases which are stated to be congenital, but I believe that such are in all instances associated with definite cerebral changes, and belong to an entirely different disease. It would be extremely difficult to distinguish between the jerky, irregular movements of an infant, and those seen in infants with meningeal hæmorrhage, or the subsequent changes induced thereby.

The ratio of cases at the Infirmary to other diseases was 1 to 180.

**Station in Life.**—The disease affects children of all grades of society. It is, however, more common among the artisan and lower classes. Our clinical information about the disease has been drawn almost exclusively from hospital work. In the Collective Investigation Report, already referred to, the returns which were furnished chiefly by general practitioners show 72·27 per cent. of cases belonging to the lower classes.

**Race.**—Chorea is rare in the negro, as shown by the inquiries instituted by Weir Mitchell some years ago among physicians in the Southern States. Sinkler, who has also written on this question, has seen but one case in the full-blooded negro. The records of the Infirmary show that no negro child of full blood has been under treatment, and only four or five cases in mulatto children are noted. As the proportion of coloured to white is in Philadelphia as 1 to 25, it would have been reasonable to expect a larger proportion of coloured patients, if chorea was as common in them as in the whites. Of 175 cases at the Johns Hopkins Hospital there were five in the negro race.

The disease is also rare among the Indians. I have made inquiries at the Indian schools, and of a number of physicians who have practised for years in the American and Canadian North-west territories. There were several references made to its occurrence in half-breeds, but not one of my correspondents mentioned a case in the full-blooded Indian. Dr. Waldron, of the Hampton Normal and Agricultural Institute, Hampton, Va., writes that he has never seen a case in an Indian, and that there had not been any in the

<sup>1</sup> *British Medical Journal*, 1887, I.



school since its foundation. Dr. McLelland, of the Lincoln Institution of Philadelphia, writes to the same effect. Dr. O. G. Given, Physician to the Indian School at Carlisle, Pa., states that during the five years in which he has had medical charge of the children there has only been one case, and that was in a scrofulous and epileptic subject. Professor Robert Bell, of the Canadian Geological Survey, who has studied the habits and diseases of the Indians from Gaspe to the Rocky Mountains, and from Lake Huron to Hudson Bay, has neither seen nor heard of a case. Chief Peter Jones, a physician who has practised for twenty years among the Mississauis and Six Nation Indians at the Reserve near Brantford, Ontario, informed Prof. Bell that he had seen three cases in half-breed girls, and had heard of two others. He does not think that it ever occurs among the full-blooded Indians. Dr. R. M. Stephen, Government Physician to the Indians of Manitoulin Island, has not met with a case in five years' practice. In twenty-five years' experience in the Canadian North-West, Dr. Schultz has seen only one case, and that was in a half-breed.

**Seasonal Relations.**—Careful studies have been made at the Infirmary for Diseases of the Nervous System upon the relative frequency of chorea at different seasons of the year, the results of which have been published by Gerhard, and by Mitchell in his well-known "Lectures on Diseases of the Nervous System," and in several important communications by Morris J. Lewis, whose latest paper, entitled "A Study of the Seasonal Relations of Chorea and Rheumatism for a Period of Fifteen Years," analyzes as regards the months of onset 717 separate attacks of chorea.<sup>1</sup> November shows the fewest attacks, namely, 24, or 3.3 per cent. A somewhat rapid rise takes place in December to 56, or 7.8 per cent. The number of cases remains almost stationary during January and February; then suddenly increases, reaching the highest point in March, namely, 101, or 14 per cent. In April a fall occurs to 63, or 8.7 per cent. Then a rise takes place in May to 80, or 11.1 per cent., the tracing after this falling gradually to its lowest point in November. Lewis' studies include a careful comparison of the number of attacks of chorea with the mean relative humidity, the mean barometer, the mean daily range of thermometer, and the amount of sunshine or cloudy weather. "The highest spring point of the chorea tracing corresponds with cool weather and a low barometer and mean relative humidity tracing, but the rise in the autumn corresponds with cool

<sup>1</sup> *Transactions of the Association of American Physicians*, Vol. VII, 1892.



weather and comparatively high barometer and mean relative humidity, the temperature record therefore giving us but little information." The conclusions drawn by Lewis from his careful studies, which have extended over a period of many years, and practically embrace all our knowledge on the subject, may here be given :—

1st.—"The seasonal relationship of chorea and rheumatism is proven."

2nd.—"There is a marked resemblance in form between the chorea and rheumatism tracings and the tracing representing the total amount of sickness present in the community per month."

3rd.—"This monthly variation in amount of sickness is not a cause in the fluctuation in the chorea and rheumatism tracing, but is itself probably due to the same influence."

4th.—"While over-study assuredly plays a most important rôle in predisposing children to chorea, the months of greatest study, and, therefore, presumably of the greatest depression of bodily vigour, do not coincide with, or even precede with any regularity, the months of greatest frequency of the disease."

5th.—"It is more than probable that 'weather' is one of the most important predisposing causes . . . although precisely which meteorological factor is the baneful one does not clearly appear. No one element of 'weather' explains fully the fluctuations of these tracings for chorea, although in the barometer and storm statistics the relationship appears to be closer than to any other etiological factor or factors that have, as yet, been advanced. It is as if a conclusion was attempted to be drawn from premises, some of which are imperfectly stated or not clearly understood, or possibly even overlooked."

6th.—"Either this apparently close relationship must be acknowledged to have an important place in the etiology of these diseases, or else the resemblance must be considered to be purely accidental, which seems most unlikely from a study of the tables shown."

**Locality.**—The disease prevails more in towns than in the country; but it is extremely widespread, as shown by the report of Isambard Owen<sup>1</sup> on the distribution of the disease in Great Britain. My friend Dr. W. N. Gordon tells me that in Exeter the number of cases admitted to the wards is proportionately very large.

**Family Disposition.**—It is not uncommon to find a special

<sup>1</sup> *Transactions of the International Medical Congress*, Washington, Vol. V, p. 157.



tendency to the disease in certain families ; thus, there were eighty cases, in which there was a history of attacks of chorea in other members of the family. Now and again we find that the mother of the child has also had chorea, and in one instance both mother and grandmother had been affected.

**Temperament.**—It is generally admitted that this plays an important predisposing rôle. Dull, heavy, phlegmatic children rarely have chorea, while high-strung, excitable, nervous children seem specially liable to the disease.

**Psychical influences.**—These have always been held to play an important part. In 86 cases, 15·5 per cent, fright was given as the exciting cause. In the majority of these no very close connection existed between the fright and the onset of the chorea, as usually an interval of two or more days had elapsed ; but in a few cases, the attack came on at once. Causes other than fright are often assigned, such as mental worry and trouble, a sudden grief, a scolding, or, in some instances, the excitement of a religious meeting. The strain in education, particularly in young girls, during the third decade, the period, to use Clouston's phrase, of "co-ordination of motion and emotion,"<sup>1</sup> is an important factor. Such phrases as "over-work at school," "worry about lessons," "examinations" occurring so frequently as they do in the records, are not without significance. Bright-eyed, intelligent, active-minded little girls from ten to fourteen, ambitious to do well at school, often stimulated in their efforts by teachers and parents, form a large contingent of the cases of chorea both of hospital and in private practice. Sturges, in particular, has called attention to this school-made chorea, as a serious injurious result of our modern methods of forced education.

**Imitation**, so often mentioned as an exciting cause, does not appear to have influenced the onset in a single case in the Infirmary series. In institutions or in a single ward, where numbers of cases have occurred in rapid succession, the disease is usually hysteria. Steiner,<sup>2</sup> who describes an outbreak of chorea minor in Prague, could however see nothing to support the imitation theory, and thought that the atmospheric influences had more to do with it. In 1880, an epidemic occurred at the Church Home for children, in Philadelphia, which is given at length in Weir Mitchell's Lectures, but in this, as in a majority of similar outbreaks, hysteria was the underlying condition. An interesting outbreak occurred at Wildbad

<sup>1</sup> *Neuroses of Development*. Edinburgh, 1891.

<sup>2</sup> *Jahrbuch für Kinderheilkunde*, 1870.



a few years ago and is described at length by Wichmann.<sup>1</sup> Some of the cases seem to have been true chorea minor, but a majority of them were examples of the rhythmic (hysterical) disorders of motion.

**Traumatic influences** have been assigned, such as falls and blows. Sinkler, in his article on chorea, in Pepper's System of Medicine, gives a case which followed shortly after a minor surgical operation. In the Infirmary records, there are a good many cases with a history of injury preceding the attack.

**Reflex irritation.**—The older writers laid much stress on so-called reflex chorea, which was thought to originate from digestive disturbances, the presence of worms—the chorée vermineuse of the French—or genital irritation. I do not find a single instance in the Infirmary records to indicate any causal relationship between these conditions and chorea.

Jacobi<sup>2</sup> has called attention to the important part played by nasopharyngeal irritation in chorea, more particularly when confined to the face, but this habit spasm, as it is more correctly called, is in reality a different affection from the true chorea minor.

Here it is worth noting that Straton<sup>3</sup> laid great stress upon the fact that erosions of the mucous membranes of the nose and throat, by affording portals of entry to micro-organisms, might be the indirect cause of chorea in children.

**Relation of Chorea minor to eye-strain.**—It has been claimed by Stevens<sup>4</sup> and others 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<sup>5</sup> as to the condition of the eyes in fifty cases of chorea in children with the following result:—Hypermetropia was present in twenty-three, or 46 per cent. ; hypermetropia in one eye, and hypermetropic astigmatism in the other in seven, or 14 per cent. ; hypermetropic astigmatism in twelve, or 24 per cent. ; myopia in one, or 2 per cent. ; myopic astigmatism in three, or 6 per cent. ; mixed astigmatism in four, or 8 per cent. De Schweinitz then adds to his report the cases reported by Stevens and C. S. Bull of New York, making a total of

<sup>1</sup> *Deutsche medicinische Wochenschrift*. 1890.

<sup>2</sup> *American Journal of the Medical Sciences*, 1886.

<sup>3</sup> *British Medical Journal*, 1885, Vol. II.

<sup>4</sup> *Transactions of the New York Academy of Medicine*, 1874-76, and *New York Medical Record*, August, 1876.

<sup>5</sup> *New York Medical Journal*, 1887, I.



227 cases, of which 112 were ametropic, and 115 emmetropic. His conclusions are expressed in the following paragraphs:—"Hypermetropia and hypermetropic astigmatism are vastly the preponderating condition in the eyes of choreic children, being found in about 77 per cent. of the cases, exactly as hypermetropic refraction is the preponderating condition in childhood, being found in 76 per cent. of the eyes of children in the elementary schools." "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." Under date of June 18th, 1894, Dr. De Schweinitz wrote to me stating that he has now examined more than a hundred cases of chorea, and he adds, "that ordinary chorea and many of the forms of facial spasm, habit spasm, etc., are materially benefited by correcting refractive errors and anomalies of the ocular muscles, just as they are helped by a variety of other treatments, but I do not believe that there is any proof to show that eye-strain of itself is responsible for their origin, with perhaps the single exception of the so-called habit spasms affecting the orbicularis and immediate facial area. Certainly many of these will disappear promptly after the refractive error is corrected without any treatment whatsoever, and they will not disappear if you do not relieve the eye-strain. In a constitution predisposed to chorea, I presume eye-strain is a very important factor in fostering and perhaps provoking attacks, but that is all."

Of the five cases of chorea submitted to Stevens by the New York Neurological Society,<sup>1</sup> not one was chorea minor, so that the statements of the report are scarcely germane to the question.

Ranney's recent report<sup>2</sup> upon this subject is of interest in two connections:—In the first place it shows the necessity of the recognition of some uniformity in deciding exactly as to the limitation of Sydenham's chorea. Of these twelve cases, nine patients were fifteen years or older, the majority of them were adults, and the muscular disorder had persisted for eighteen months, the shortest period, to thirty-one years. I do not think that any one of the twelve cases could be really called true Sydenham's chorea, but that nearly all were one or other of the varieties of habit spasm or tic, some possibly of hysterical chorea. The second point is the enormous importance of careful examination of the eyes in these chronic forms of disordered *musculature*, to use Roth's term.

<sup>1</sup> *Journal of Nervous and Mental Diseases*, 1889, p. 649.

<sup>2</sup> *New York Medical Record*, May 5th and 12th, 1894.



I know that there are circles in New York in which Dr. Ranney bears the reproaches of a prophet, but—*litera scripta manet*.

**Arthritis (Rheumatism).**—The association of arthritis and chorea did not escape the observation of physicians in the last century, and Bouteille<sup>1</sup> gives in his monograph two observations of Stahl, and two of Sauvages. Bright states that in the edition of the "Syllabus, or Outlines of Lectures on the Practice of Medicine," published at Guy's Hospital in 1802, rheumatism was distinctly recognized as one of the causes of chorea, and he says also that in the later editions, as in that of 1820, it was stated that "chorea sometimes alternates with acute rheumatism." Bouteille makes the rheumatic one of the minor sub-divisions of his deuteropathic or secondary chorea. So far as I can see he lays no stress on it in connection with the essential or primary form. Copeland<sup>2</sup> writes:—"The association of this disease with rheumatism has been observed by me on several occasions, and in nearly all there has been a marked disposition of the rheumatic affection to recede from the joints or extremities, and attack the internal fibro-serous membranes, as those of the cerebro-spinal axis, and the pericardium." J. C. Pritchard,<sup>3</sup> of Bristol, reports several severe and fatal cases of chorea, and speaking of one of them in which pericarditis was present, he says: "the disease seemed to have arisen from the metastases of rheumatism." The association between chorea and rheumatism was evidently very clearly recognized by the Guy's Hospital physicians during the early decades of this century, and Bright<sup>4</sup> has directed particular attention to this question; but in Babington's paper on Chorea in the Reports,<sup>5</sup> no special stress is laid upon rheumatism. He remarks that "rheumatism also, when it affects the heart and pericardium, may give rise to the disease through the irritation of the plexus and ganglia, which so entirely surround that organ, and the origin of its great vessels." Oddly enough, considering that his paper was written only a couple of years after Bright's communication, he gives the credit of his discovery of the association of disease of the heart with chorea to Addison, stating that "should further investigation prove chorea to be more intimately dependent on disease of the heart or pericardium, than has been

<sup>1</sup> *Opus cit.*, p. 291.

<sup>2</sup> *London Medical Repository*, Vol. XV.

<sup>3</sup> *London Medical Repository*, Vol. XXI, 1824.

<sup>4</sup> In Vol. II. of his *Medical Reports*, and in *Medico-Chirurgical Society's Transactions*, Vol. XXII, 1839.

<sup>5</sup> Vol. VI, 1841.



hitherto supposed, the merit of the discovery will certainly be due to Dr. Addison."

The strongest support to the rheumatic theory of the origin of chorea was offered by the French writers, particularly by Germain Sée<sup>1</sup> in 1850, and by Roger<sup>2</sup> in 1866. Sée's conclusions may be briefly expressed as follows:—Of two rheumatic infants at least one will be choreic; and of five choreic children there are two rheumatic; while Roger concluded that articular rheumatism, chorea, and heart disease (endocarditis) were three terms of one and the same pathological state or phase—*la chorée rhumato-cardiaque*. Since the appearance of these papers every writer on the subject has dealt more or less fully with the question of the relation of the two disorders.<sup>3</sup> The statistical method which has been applied to the solution of the problem has served to bring out very striking discrepancies. The German writers, as a rule, have not placed very much stress upon rheumatic relations of the disease. Thus Steiner, of Prague, found only four instances of acute arthritis in 252 cases of chorea. English authors place the coincidence very much higher, and make it from 20 to 50, and even 70 per cent.

So far as the statistics are of any value, the following statements may be made with reference to the relations of arthritis and chorea in the United States. Of the 554 cases at the Infirmary for Diseases of the Nervous System, Philadelphia, in 15.5 per cent. there was a history of rheumatism in the family; eighty-eight cases, 15.8 per cent, had had at some time or other, either prior to, with, or subsequent to the attack of chorea, acute or subacute articular swellings. In thirty-three cases there was a definite history of pains in various parts, sometimes described as rheumatic, but not associated with joint trouble, and if these are regarded as rheumatic, and added to those with manifest arthritis, the percentage is raised to nearly 21. At the Johns Hopkins Hospital, of 175 cases treated in the wards and in the Dispensary to January 1st, 1893, there was a history of arthritis, acute or subacute, in twenty-seven cases, or in 18.24 per cent.

At the discussion which took place before the American Pediatric Society at Boston,<sup>4</sup> Charles W. Townsend reported 148 cases with a

<sup>1</sup> *Mémoires de l'Académie de Médecine*, Tome XV, 1850.

<sup>2</sup> *Arch. Gén. de Médecine*, December, 1866, and January, 1867.

<sup>3</sup> In any discussion on this point, arthritis and rheumatism must be regarded as convertible terms, though the joint affections of chorea may be no more *rheumatic* than those of cerebro-spinal meningitis.

<sup>4</sup> *Transactions of the American Pediatric Society*, Vol. IV, 1892.



history of rheumatism in thirty-one, or 21 per cent. ; Crandall, eighty-eight cases with rheumatism in some form, either before or after the chorea, in 54 per cent. ; M. Allen Starr, 385 cases, with a history of rheumatism in 18 per cent., growing pains or pain of an indefinite kind not being included under this head. In the discussion which followed, Jacobi took strong grounds in favour of the close relationship of the two affections, an opinion supported strongly also by Holt. No unanimity of opinion was reached on the question ; the only very high rheumatic percentage was that given by Crandall. Of course, in making up statistics, unless the histories have been taken with actual reference to the points under discussion, not much reliance can be placed upon them, and it is this very fact which makes the figures from the Infirmary for Diseases of the Nervous System of special value, inasmuch as the chorea histories have been taken in special books provided with full and detailed printed questions, particularly on the possible rheumatic manifestations. Of the last 144 cases of the Infirmary series, in almost every one of which I myself made the most careful inquiries of the relatives as to rheumatic features, there were only twenty-five with articular pains or swelling, and only six of these had had acute inflammation of joints. In England, among the more recent statistical investigations may be mentioned the report of the Committee on Collective Investigation of the British Medical Association, which gives, out of a total of 439 cases, ninety-seven with a rheumatic history, a little more than 22 per cent., with which the largest collected American statistics closely agree. Of the seventy-three fatal cases collected by me (see Appendix) twenty-nine are stated to have had rheumatism, three had the subacute form, in four it was not definitely stated, and thirty-seven had not had acute arthritis.

As insisted upon, especially by Sée and by Roger, the arthritis in a very large proportion of all cases precedes the chorea, which develops with its subsidence, or may not follow until convalescence has been well established. In other instances the chorea precedes the rheumatism. This, however, is rare. In the report of the Collective Investigation of the British Medical Association on rheumatism, analyzed by Whipham, chorea was found to have preceded rheumatism in less than 2 per cent. of the cases.

The manifestations of rheumatism in childhood are extremely varied, and often so slight that they are readily overlooked. I well remember, in 1872, at the University Hospital, a bedside talk of



Sir William Jenner's on a case of a little girl with endo-pericarditis, and only the most trifling articular manifestations, which had been entirely overlooked before her admission. The remarks made an indelible impression upon me, and I have been in the habit of seeking carefully for the minor manifestations of the disease. The London physicians have done much to broaden the conception of rheumatism in childhood, of which subject Cheadle's<sup>1</sup> lectures give a full presentation.

Without having the facts upon which to base a positive statement, I am of the belief that rheumatic fever is not nearly so frequent in Philadelphia and Baltimore as in London. Here, at any rate, some of the most striking rheumatic manifestations are conspicuous by their absence; for instance, a case of subcutaneous fibroid nodules is a great rarity.<sup>2</sup> In my five years' service at the Infirmary for Diseases of the Nervous System not a single instance was seen. Since 1881, when I saw cases at the Great Ormond Street Hospital, I have been in the habit of looking for them in a case of rheumatism as systematically as I examine into the condition of the heart. I have seen a larger number of instances in adults than in children. The matter of their rarity was a subject of comment among the members of the Pediatric Society at the Boston meeting in 1892. They may exist independently of acute arthritis, or even of any rheumatic manifestations. I have had in my wards for three years a girl with chronic valve disease—mitral and tricuspid—who has never had, so far as can be ascertained, any signs of rheumatism. During the past eighteen months she has developed many subcutaneous fibroid nodules about the elbows and hands, and along the tendons in various places. As I write (June 30, 1894), there has been admitted to the wards the first case which I have seen in this country with the combination of chorea, arthritis, subcutaneous fibroid nodules, and mitral endocarditis. The only case which, so far as I know, has been reported in the United States with this combination is the remarkable one by C. H. Brown.<sup>3</sup>

**Infectious Diseases.**—With reference to its development in, or as a sequence of, other diseases the following statement may be made. Scarlet fever with arthritis may be a direct antecedent.

<sup>1</sup> *The Rheumatic State in Childhood*. London, 1889.

<sup>2</sup> So far as I know the first case reported in the United States was from my clinic at the University Hospital, Philadelphia, by Dr. J. K. Mitchell (*Univ. Med. Magazine*, Vol. I.)

<sup>3</sup> *Journal of Nervous and Mental Diseases*, 1893.



Chorea developed in three of 533 cases of scarlet fever reported by Carslaw.<sup>1</sup> A previous history of this disease was obtained in 141 of the Infirmary cases, but in not one was the sequence immediate. Sturges states that a history of previous whooping-cough is met with very frequently in choreic children. The disease may develop acutely in the course of gonorrhœa, secondary syphilis, and in septic infections, such as puerperal fever, pyæmia, and multiple suppurative polyarthritis. In Litten's important paper<sup>2</sup> full details are given of cases associated with these acute infections. Cases have also been reported as occurring after diphtheria, measles, scarlet fever, and typhoid fever. Kinnicutt and others have reported instances of the association of chorea with malaria, a connection probably accidental and not causal, since chorea does not prevail to a greater degree in malarial regions. With the exception of acute polyarthritis (rheumatic fever) and certain forms of septicæmia there is no very intimate relationship between chorea minor and the infective disorders. Canine chorea also follows an acute infectious disease of the dog.

On the other hand the acute exanthems developing in the course of chorea usually check the disease (Rilliez and Barthez). On this point Radcliffe's remarks are worth quoting: "At any rate, there are many cases on record of measles, scarlet fever, rheumatic fever, or some other fever, being developed during the course of chorea, and in which the choreic symptoms have been suspended during the fever. I have met with seven such cases. Indeed, as far as I have had the opportunity of judging, the constant rule appears to be, that the chorea is aggravated in the initial stage of the fever—that is, in the cold stage, or stage of irritation—and suspended more or less completely when the stage of reaction, or hot stage, is established; and that, in relation to rheumatic fever, the place of chorea is either before the fever (often a long time before) or after the fever (often a long time after);"<sup>3</sup> and he quotes the maxim of Hippocrates, "febris accedens solvit spasmos."

S. West reports the case<sup>4</sup> of a child, aged ten, with chorea, in whom the onset of typhoid fever checked the movements. Trousseau also reports illustrative cases.

**Other Diseases.**—Anæmia, upon which as a causal factor much stress has been laid, appears to be more often a sequence than an

<sup>1</sup> *Glasgow Medical Journal*, 1891.

<sup>2</sup> *Charité Annalen*, Bd. XI.

<sup>3</sup> *Reynolds' System of Medicine*. Article Chorea.

<sup>4</sup> *St. Bartholomew's Hospital Reports*, Vol. XXII.



antecedent, and though cases do develop in children who are anæmic and in poor health, this is by no means the rule, and my experience lends no support to the view warmly advocated, among others by Rachford, that an impoverished blood condition plays an important rôle in the causation of the disease. I have, however, seen several instances in which chorea developed in chloritic girls at puberty.

Litten<sup>1</sup> quotes a case of Roeser in which chorea developed six days after a copious bleeding at the nose, and proved fatal. Fresh endocarditis was found at autopsy. He speaks also, but gives no details, of two cases of chorea developing in the course of pernicious anæmia, both of which ended fatally, and presented at the autopsy signs of a fresh endocarditis.

**The Relations of Chorea and Hysteria.**—In the first place, it is not uncommon to see hysterical manifestations in young girls with chorea, which are simply complications, and mean nothing more than that the disease has attacked an hysterical subject, so that we have a combination of the two affections. Many such instances are to be found in our records, but as a rule the hysterical symptoms were quite subsidiary.

Secondly, it is quite possible for hysteria to actually simulate chorea minor. Such instances are very exceptional. A case was reported by Debove to the Société Médicale des Hôpitaux in 1890. The man, aged 21, had all the features of chorea minor; the movements were wide-spread and excessive. Well-marked hysterogenic zones were determined. It was decided to try compression over the parts of the abdomen corresponding to the ovarian regions in women, and as a matter of fact the strongest possible pressure at these points caused the immediate cessation of the movements. The patient had also anæsthesia of the pharynx. The movements began the day after he had had a serious trouble with his *fiancée* and an attempt to commit suicide, in which, however, the rope broke.

Thirdly, there is a possibility that chorea minor and the rhythmical or true hysterical chorea may coexist in the same person. Such an instance was reported by Séglas to the Société Médicale des Hôpitaux in 1891. The patient was a girl aged 15, who had had ordinary choreiform movements, at first of the face and of the left side, making continual gesticulations and incessant grimaces, and becoming worse, so that she was unable to speak and had all the characteristic features of chorea minor. Within two months, after

<sup>1</sup> *Op. cit.*



some alternations of improvement and recurrence, the choreic movements improved and gradually disappeared from the face and from the limbs. In the left arm they continued, but no longer presented the characteristic inco-ordination which had formerly been present. These gradually increased until there were marked rhythmical movements of the arm of the so-called "hammer" type—*chorée malléatoire*. She had also at intervals attacks of laughing and crying. There was hyperæsthesia in the ovarian regions, and she became excessively emotional.

A series of thirty-four cases of chorea with hysteria have been collected by Duchateau (Paris Thesis, 1893).

**Poisons.**—There are a few observations pointing to the occurrence of chorea as a sequence of poisoning by gases or medicines; thus *Observation IV.* in Leudet's<sup>1</sup> paper, a man aged 61, an alcoholic, who had frequently made attempts at suicide, was found asphyxiated by the vapour of charcoal. He had on admission movements of flexion and extension of the right arm, quite forcible and marked. After recovery of consciousness, on the two successive days the movements persisted. There was no paralysis. They ceased on the 12th. This case, which is often quoted, has of course nothing to do with chorea; the movements were such as is now well known not infrequently occur in poisoning by gases.

Demme's case<sup>2</sup> is more probable: a boy, aged six, with caries of the cervical vertebræ, had an iodoform bougie placed in the fistula on August 1st. He did not feel very well after it for a week or so. On the 18th a second was inserted, and on the following morning choreic movements began in the arms and legs. He gradually improved, and by September 15th was well. About the end of October the fistula was injected with iodoform again, and four times subsequently. The choreic movements returned.

<sup>1</sup> *Archives Générales de Médecine*, May, 1865.

<sup>2</sup> Abstract in Schmidt's *Jahrbücher*, Bd. CCXXIV.



## CHAPTER II.

## SYMPTOMS.

Symptoms—Mild, severe, and maniacal forms — Chorea Insaniens — Special Symptoms—Motor disturbances—Muscular weakness.

ALTHOUGH the disease may begin quite abruptly, as a rule the cases present premonitory symptoms, motor or psychical. The child is restless and unable to sit still, a condition well expressed by the term "fidgets," often used by the mothers in describing the onset. The entire disposition may be changed, and the child becomes irritable, cross and unmanageable. Emotional disturbances are common, the child crying on the slightest provocation. Night terrors have been noted in many instances. Punishment inflicted at this stage by thoughtless parents or teachers aggravates the mischief, or even precipitates the attack. The appetite may fail, and anæmia not infrequently develops. Headache is a not uncommon complaint. Less frequently there may be at this stage pains in the limbs, usually in the legs, or about the joints. After a week or more of these symptoms the restlessness increases, and the sudden involuntary movements characteristic of the disease begin. These are often first noticed at table, when the child spills a tumbler of water by a sudden jerk, or upsets a plate, or drops something. Beginning as a rule in the hands and arms, the brusque, involuntary movements give a very characteristic stamp to the muscular disorder. In any large series three groups of cases may be recognized; the *mild*, in which the affection of the muscles is slight, the speech not seriously involved, and the general health not much disturbed; the *severe*, in which the movements are general, the power of speech is lost, and the patient is unable to get about and help himself; and the *maniacal* form, *chorea insaniens*, characterized by profound cerebral disturbances.

**Mild Chorea.**—A very large proportion of all the cases belong to this group. The symptoms are annoying, and perhaps distressing, but never alarming, and the child is always able to walk about



and to talk. Sometimes, however, with very slight motor symptoms the psychical features may be marked, showing themselves not often in mania, but in mental depression or even melancholia. The following case is a good illustration :—

*Acute Rheumatism, followed by Chorea, Second Attack the following Year; Admission in Third Attack, no Heart Complication; Rapid Recovery.*

Ida A., aged seven, was admitted to hospital April 24th, 1887, in her third attack of chorea. The mother has rheumatism. Early in January, 1885, the child had an attack of acute rheumatism; was in bed four weeks. One month after convalescence the chorea began, and for several months she twitched, chiefly on the right side. On January 29th, 1886, she had a second attack, and attended at the clinic for three months. At this time there was no rheumatism, and the heart-sounds were clear. About the end of March of this year, her mother noticed that she was extremely nervous and irritable, and on the 30th she had irregular jerky movements of both hands and of the shoulders, with an occasional facial grimace. On April 20th she returned very much worse; the movements of the arms constant, speech not affected. She was admitted to hospital, put to bed, given Fowler's solution,  $\mathfrak{m}$  v, t. i. d., galvanism to the spine, and massage of the limbs daily, and ordered to be kept quiet and undisturbed. There was no heart murmur. She improved rapidly, and at the end of a week the movements had almost ceased. On May 7th she was discharged well.

The case is a very characteristic one. A child of a family with a rheumatic history, had an attack of rheumatic fever, during convalescence from which choreic movements began, and continued for several months; then came the spring recurrence in two successive years, which is so marked a feature in many cases.

**Severe Chorea.**—Cases belonging to this category are very much less common, but any mild attack in which the child is able to get about and to talk plainly, may develop into the severer form, in which the muscles are so involved that volitional movements are impossible, and the power of speech is lost. No more distressing disorder of childhood is to be seen than this severe type of chorea. Fortunately the cases are not very common, and in children the disease does not often pass into the most intense grade, in which there is active mania and high fever.

The following is a good illustrative case :—



*History of Fright ; General Chorea, Movements Violent and Incessant, no Heart Complication.*

Ella A. (sister of the preceding), aged nine. Has not had rheumatism. Has been nervous at school. Failed in some lessons. Was frightened at a lecture at the Wagner Institute, and three months ago was much upset by seeing a child injured and bleeding. The twitching was first noticed in right arm. When brought to clinic, December 30th, she presented the appearance of a rosy, plump child. The condition was noted as follows :—Choreic movements are universal ; the hands and feet jerk about in a most violent manner, the movements usually beginning in the left side, and extending to the right. They number from thirty to thirty-two in a minute, and are so strong that the most powerful restraint fails to repress them entirely. The hands are drawn from the lap violently upward, striking the chest, or they rise suddenly and fall heavily on the knees. When she is seated on a chair the feet are lifted from the floor and come down suddenly with a loud stamp. The face is in constant movement ; speech is almost impossible, and she presents a truly pitiable condition. So violent are the movements that she has frequently injured and bruised herself, and she has several times inadvertently struck her mother. She is intelligent, sleeps well, even soundly, the movements ceasing completely. It was impossible to examine the heart on account of the extreme jactitation. She took Fowler's solution, up to  $\text{m xv}$ , t. i. d., with benefit ; but the case was protracted, and although the violent movements subsided, it was several months before they disappeared.

Such cases are often carried into the clinic, in some instances covered with bruises on the exposed and prominent portions of the body.

**Chorea Insaniens.**—No stronger point in favour of the existence of a distinct and essential malady, chorea, is to be found than the occurrence of cases of great severity, which represent, so to speak, the malignant manifestations of the disease, comparable to those met with in other acute affections. Just as we have instances of very mild scarlet fever with trifling symptoms, and all grades of the disorder up to the scarlatina maligna, so in any large series of cases of chorea, the grades of the affection pass insensibly from the milder, trifling forms to types of such intensity that they equal in severity the more aggravated forms of acute nervous disorders, such as tetanus, hydrophobia, and cerebro-spinal fever. Maniacal chorea



—the chorea insaniens of Bernt—is a truly terrible affection. The following are the only two cases which I have seen :—

*History of Fright, Rapid Onset of Chorea, Delirium, Fever, Death on the Tenth Day, Acute Endocarditis.*

Emma M., a strong girl, aged eighteen, was admitted to the Montreal General Hospital, October 17th.<sup>1</sup> She was a waitress at an hotel, and when carrying a tray was badly frightened by two men who were fighting, and dropped her dishes. A severe reprimand only increased her fright. The next day she packed her trunk and went home, a distance of thirty miles. On the way she met her father, who insisted that she should return to Montreal. By this time her hands and arms were in constant motion. On the way back her father got drunk, and threatened to punish her if she did not keep quiet. She was admitted five days after the first fright. The arms and legs were in constant motion, jerking about in all directions ; the face was also affected. She was rational, but could scarcely speak. On the night of the 19th she did not sleep, but raved and talked all the time ; the movements were incessant. On the 20th, 21st and 22nd, this condition persisted, or rather grew worse ; the temperature ranged from 101° to 103° ; the tongue was dry and cracked, and she became much exhausted. On the night of the 22nd the temperature rose to 105°, and she died five days after admission, and ten days from the onset of the symptoms. The autopsy showed recent soft, warty vegetations on the auricular surface of the mitral segments. (See Morbid Anatomy.)

*Rheumatism at 16 and 23, the last Attack followed by Chorea. Onset without known cause of General Chorea, Admission at end of First Week. Fever, Skin Rash, probably Secondary Syphilide. Delirium, Parotitis, Cyanosis, Exhaustion and Death. Endocarditis, Broncho-pneumonia.*

Miss A., aged 27, a teacher by occupation, was admitted to the Johns Hopkins Hospital on December 24th, 1889, with irregular movements of the extremities and body.

*Family History.*—The patient has three brothers and three sisters living, and in good health. A brother died of pneumonia when young ; he had chorea for a month at the age of ten years.

<sup>1</sup> Case reported by Dr. Geo. Ross, *Canada Medical and Surgical Journal*, Vol. XI.



Her grand-parents lived to an advanced age. There is no further family history of import.

*Previous History.*—She has been generally strong and healthy ; had scarlet fever, diphtheria and measles, when a child. Menstruated at the age of sixteen. About eleven years ago she had an attack of rheumatism, lasting four weeks ; the shoulders and hands were then swollen. She recovered her health completely afterwards. Since that time she has been teaching, and studying excessively hard, so much so that at times her friends feared she would lose her reason under the mental strain. A second attack of rheumatism occurred four years ago, after a period of intense application to her studies. The hands were much swollen, the other joints slightly so ; all were exquisitely tender. She had been ill some days, when, owing to some excitement, her mind became affected, and for five weeks she was delirious. At this time she was supposed to be suffering from meningitis. During three of these weeks she had the same irregular movements of the legs and arms as in the present illness, though they were not so violent. These muscular jerkings and mental symptoms disappeared, and she continued in good health until August, 1889, when the headaches, which had been troubling her slightly during the previous winter, became more severe. The eyes now often pained her, so that she could not read for any length of time. The back also felt weak after very slight exertion, and she became very nervous. Shortly afterwards she was confined to bed for three weeks, at the end of which time she went away on a visit. During this visit she was able to go about, ride and dance, though still suffering from headache at times. During the night of December 17th, 1889, she was awakened by a jerking of the muscles of the right arm ; then those of the other arm and finally the legs, face, and body became involved. This condition has continued unchanged until admission.

*Present State.*—She is unable to sleep. The mind is clear. Articulation is difficult. She tosses continually from one side of the bed to the other ; the movements are very violent. Nutrition is good, as is also the colour of the skin. There are well-marked and very varied choreic movements, affecting all parts of the body. On the face and arms, here and there on the legs, and sparingly on the trunk, there is a flat, papular, bronze-coloured rash. In places it is distinctly scaly. It is most abundant on the inner side of the wrists, on the palms of hands and bends of elbows. It is symmetrical. The cervical glands are enlarged, hard, and nodular ; there is slight



enlargement of the inguinal glands on the left side. The pharynx is ulcerated; uvula healthy. Careful examination of the fauces was not possible on account of the movements of the head. There is some patchy alopecia. The heart-sounds are clear, both at apex and base; the apex beat is in the fifth intercostal space. On the sides and under surface of the clitoris is an elevated and excoriated, non-indurated sore. A further examination of the vagina and cervix was not possible. There is no incontinence of urine or of fæces.

*December 28th.*—The patient was very restless up to midnight, and then, frightened by an imaginary something, she jumped out of bed, shrieked wildly, and apparently did not recognize the nurse. She was afterwards quiet for a few minutes at a time, but had periods of maniacal excitement, starting at every sound. The muscular movements are not so exaggerated as during the previous night, but the mental condition is decidedly worse. At 3.50 A.M. chloralamide, grs. xxx, was administered, and she became more quiet, and at 4.30 A.M. was sleeping heavily. The choreic movements ceased during deep sleep.

*December 29th.*—Patient is somewhat better this morning. The muscular contractions are not so violent. The heart-sounds are clear, both at apex and base. There is no joint tenderness. Several hypodermic injections of morphia were given during the evening; under its influence she slept from 9 P.M. until 1 A.M. Was then restless until after 3 A.M., when she slept for three-quarters of an hour.

*December 30th.*—The choreic movements are quite general this morning; the left arm, which was quiet yesterday, is now tossed about equally with the right. There is less mental confusion—patient answering questions more rationally. The tongue is moist and furred. Takes nourishment better than yesterday. Under the influence of a third of a grain of morphia, she slept uninterruptedly through the night.

*December 31st.*—Is very restless this morning. The choreic movements are about the same as yesterday. Pupils normal. She speaks very volubly and irregularly; speech is not so distinct as previously. The tongue is moderately dry. The pulse is 144, not especially weak. She still takes nourishment very well by the mouth. There is a very marked friction erythema on the arms, face, feet, and knees.

*January 1st.*—Patient slept more than six hours last night under



the influence of morphia, given at 9.30 P.M. Pulse 100, very irregular. Seemed better this morning; appetite good. Muscular twitchings not so violent. At mid-day she fell out of bed, and when picked up was found to have passed a copious liquid stool. After removal to bed she became somewhat cyanosed. Pulse 132, extremely feeble. Ether was given hypodermically, and brandy and hot water by mouth and rectum. At 5.30 P.M. the pulse was still rapid, but of better volume. She is still restless, and has occasional spells of shrieking.

*January 3rd.*—Since the last note was taken patient has been in a condition of stupor, lying on back and breathing regularly, twenty-four to the minute. Pulse 120, not decidedly better than two days ago. There are occasional choreic movements, limited to the arms. She had several watery stools this forenoon. The abdomen is soft. The tongue and lips are dry and brown. At the angle of the jaw on the right side there is a hard and tender swelling over which the skin is reddened. The heart-sounds at apex and base are clear. There are no choreic movements.

*January 4th.*—The abdomen is soft and a little distended; there are no spots visible on it. Respiration rapid, forty-four to the minute. Fine crepitant râles are present in the left axillary region, with an expiratory rub. There is no heart murmur. The same fine crepitant râles are present in right axillary region, also with an expiratory rub. The patient sank gradually during the day, death occurring at 6.30 P.M.

Autopsy, by Dr. Councilman, fourteen hours after death.

Body of medium size, 157 centimetres long, slightly built, well nourished. Posterior surface much congested. Right side of face swollen, reddened and œdematous; a considerable amount of fluid blood flowed from the ear. Bloody fluid also escaped from the nostrils. On the anterior surface of the right thigh, and on the internal surface of the left near the knee, are depressed cicatrices. Here and there over the body, generally corresponding to creases in the clothing, are congested areas.

*Cranium.*—Scalp thin and firmly adherent. Skull of ordinary thickness, symmetrical. External surface of dura mater smooth. Fluid blood in longitudinal sinus. Dura slightly attached to bone everywhere. The brain with the meninges weighed 1,274 grammes. The lateral sinuses are free from clots. The petrous bone of the right side is hyperæmic. Pia mater is slightly œdematous. The arteries at the bases are normal. No abnormal change is to be seen



in the cerebral tissues. There is no marked hyperæmia of the cortex; no hæmorrhages.

*Abdominal Cavity.*—The subcutaneous fat is slightly developed. Muscles red. Peritoneum smooth. The cavity dry. The liver and spleen are free from adhesions. The liver, spleen and pancreas congested. The kidneys showed foetal lobulation.

In Douglas's *cul-de-sac* are numerous ecchymoses. The uterus is large, blood-vessels prominent, the tissue hyperæmic. Small corpus luteum in left ovary. Immediately around the meatus urinarius, the mucous membrane is slightly swollen. There are no ulcerations or cicatrices about the genitalia.

*Thoracic Cavity.*—Præcordial space uncovered by lung tissue small. Both layers of the pericardium are smooth. The heart is contracted. In the right cavity a small amount of fluid blood; same on left side. Heart flesh pale and flabby. On the auricular surface of the mitral valve are a few fresh warty vegetations. Columnæ carneæ in left ventricle fibrous. Coronary arteries normal. The lungs are free from adhesions. In the upper part of the left lung is an area of consolidation, the pleura over it being covered with a slight fibrous exudation. On section two foci are seen, the centre of one of them being somewhat whitish. Adjoining them is a depression on the surface of the lung, and beneath this is a caseous focus 3 millimetres in diameter, surrounded by indurated lung substance, the induration extending downward into the lung. The lung tissue in the lower lobe is hyperæmic. Pus can be squeezed from all the small bronchi, and there are small scattered foci of consolidation. On the upper border of the lower lobe is a small recent infarction. The surface of the lower lobe of the right lung is hyperæmic. In the upper lobe are numerous areas of consolidation, the largest about 2 centimetres in diameter. In the lower lobe pus can be squeezed from all small bronchi. There are also numerous small foci of consolidation. The mucous membrane of the bronchi of both lungs is intensely hyperæmic.

The right parotid gland presents extensive purulent infiltration. The external ear is intensely reddened and hyperæmic.

*Pharynx.*—The mucous membrane of the pharynx is swollen and intensely hyperæmic.

*Spinal Cord.*—The membranes and medulla are apparently normal.

Microscopic examination of the fresh tissue by Dr. Councilman showed the heart muscle to be slightly degenerated (fatty), the



epithelium of the kidneys swollen and granular, fatty in the upper collecting tubes. The liver was normal. Cultures were made from the blood in the right and left ventricles, and from the valvular vegetations, also from the parotid gland. Those from the right ventricle remained sterile, probably on account of overheating; from the others there was an abundant growth of the *staphylococcus pyogenes aureus*.

*Anatomical Diagnosis.*—Acute endocarditis of mitral valve. Abscess of parotid. Catarrhal pneumonia of both lungs with bronchitis. Old tuberculosis of the left lung.

This patient was febrile throughout. Unfortunately the temperature sheet from December 24th to January 2nd was lost. On the morning of the 2nd in the rectum the thermometer registered  $105.2^{\circ}$ . It fell on the morning of the 3rd to below normal; rose throughout the 4th, and was  $104^{\circ}$  before her death.

Chorea insaniens is thus defined by Bernt<sup>1</sup>:—"Chorea insaniens, quando affecti simul insaniae specie tentantur, variosque et nonnunquam truculentos, sibi inconsuetos mores, exhibent."

A very large proportion of all the fatal cases of chorea have active delirium with fever. The subjects of the maniacal form are commonly young females, between the ages of fifteen and twenty; but children are also attacked, and one of the most rapidly fatal cases on record (No. 15,<sup>2</sup> Cook and Beale) in which death occurred in 130 hours, was in a girl of nine years of age. The disease may be severe from the outset, but more commonly the case begins as one of ordinary chorea, and the special symptoms develop gradually. So predominant are the mental symptoms that cases are not infrequently admitted to Asylums (Nos. 20 and 21). At the outset there may be hallucinations, which quickly give place to a chattering incoherency, less often a furious mania. It is to be noted that, as in the case just given, the movements may diminish greatly in intensity or cease altogether before death. Fever is an almost constant, but not an invariable, accompaniment. The temperature may be very high; in No. 18 of the fatal cases it reached  $107.6^{\circ}$ , in No. 11,  $107^{\circ}$ , and in No. 16,  $41.5^{\circ}$ . The duration of the fatal cases is rarely more than two weeks. Though a very pernicious form, the mortality is probably not so high as the literature would indicate, since there is a natural tendency to report only the more severe

<sup>1</sup> *Opus cit.*, p. 21.

<sup>2</sup> These numbers refer to the tables of fatal cases in Appendix.



cases. Gee lost only one out of seven ; and one of his patients with high fever and double parotitis recovered.<sup>1</sup>

**Motor Symptoms.**—The various muscle groups are not equally affected ; in a majority of the cases the hands are first involved, then the face, and subsequently the legs. The movements begin most often on the right side ; thus of 410 cases analyzed with reference to this point, there were 126 cases in which they were at first dextral, starting usually in the hand, and only 80 instances in which the left side was first attacked. The movements rarely begin in the legs ; in only six instances out of the series of 410 cases was it so noted. In about a dozen instances the movements were said to have been general from the first, but the fact that early symptoms are readily overlooked must be taken into consideration. In 126 of the cases the movements became general. The muscles of the trunk and thighs are as a rule bilaterally affected. We rarely see the one group of shoulder muscles involved without the other, and the facial grimaces are not often confined to one side.

Weir Mitchell has pointed out that there are differences in the character of the irregular movements ; thus, in one group comprising the greater number of cases there was “awkwardness and inco-ordination of voluntary movements, followed soon or late by automatic or unwilled clonic spasms of various parts.” In a second group the disease does not get beyond the state of inco-ordination, and irregular movements only occur during willed actions. In a third group there are “constant, automatic, irregular, clonic spasms, usually of the hands, but during volitional acts these entirely vanish, and the most complicated acts are well performed without obvious inco-ordination.” In rare instances the movements become much less marked, or even cease when the patient stands up (Bernt).

In nearly one-fourth of the cases the speech is affected. When slight, the disturbance may be limited to a slight impairment or hesitancy, and the child utters the words plainly ; in other instances speech is an incoherent jumble ; the tongue and lips move, and sounds are uttered ; but it is impossible to recognize the words. Sometimes a few words are spluttered out in an explosive manner. The inability is in articulation rather than in phonation. The lips and tongue are more concerned than the intrinsic laryngeal muscles, or those of the expiratory group. In one case, however, with marked spasm of the inspiratory muscles and involvement of the

<sup>1</sup> Bernt says, p. 77, “*Aegre etiam Chorea insaniens curam admittit.*”



larynx, as shown by the whistling inspiration, the speech was not at all involved. In rare instances a difficulty in articulation is the first symptom noticed. As a rule, when the involuntary movements are of a very severe grade, the child will make no attempt to speak. It is to be remembered also that with very slight motor phenomena, and even without involvement of the facial muscles, the child may not talk at all, owing to psychical disturbances.

As a rule the movements cease during sleep, but in some cases they persist. Possibly in some of these instances the mothers have mistaken the irregular twitching, so often seen during sleep in nervous children, for the movements of chorea. In Case 121 of the Infirmary Series the movements are said to have been first seen while the child was asleep, and in Case 119 (*b*), in a very severe attack, the statement as to the persistence of the movements during sleep is very specific. Indeed I have known movements to occur in a choreic child, even while asleep, of such severity that it required the nurse to keep it in bed; thus in Med., No. 97 (Johns Hopkins Hospital), Charles I., aged 12, admitted in a fourth attack of very great severity, it is noted that on three separate occasions during the night of the 27th of June he had severe attacks of muscular movement in his sleep, which were very violent, and caused him to toss about. It is also noted that while asleep his arm and leg moved almost incessantly.

Do the choreic movements extend to the muscles of organic life? The great gastro-intestinal muscle is never involved; there are certainly no symptoms which can be referred to irregular contractions of the coats of the stomach or bowels. The patients rarely complain of colicky pains. The sphincter ani is uninvolved, and the bladder acts normally as a rule, even in aggravated cases. Naturally there are a few instances of incontinence of urine, but not more than one would expect in the records of any other disorder in children. Spasm of the muscle fibres of the bronchi, inducing asthmatic attacks, is not met with even in those cases in which the respiratory muscles are involved. The irregularity and rapid action of the heart, as well as the variability of the mitral systolic murmur, have been thought to be due to disturbed rhythm in the ventricular contractions, and to choreic spasm of the papillary muscles, but for this there is no very satisfactory evidence. The point will be referred to again under the cardiac symptoms of the disease. Braxton Hicks has reported a case<sup>1</sup> of chorea in pregnancy, in which during the attack the usual orderly contractions of the uterus became very

<sup>1</sup> *Lancet*, 1889, II.



irregular. The pupils may be dilated or insensible to light ; sometimes they are unequal.

**Muscular Weakness** is a not uncommon symptom.

Sydenham spoke of the "unsteady movement of one of the legs which the patient drags." Dover and Mead both regarded chorea as a paralytic affection ; while Bouteille in the first case given in his monograph describes a paralytic enfeeblement of the affected side. Todd<sup>1</sup> has described very fully the paralytic form, to which West has given the name limp-chorea (chorea mollis).

The condition is usually one of enfeeblement of the muscular strength, rarely of actual paralysis. The distribution of the weakness may be hemiplegic, paraplegic, or more frequently monoplegic. In twenty-eight of the Infirmary cases in which there was a special note of loss of power, the distribution was as follows :—paraplegic, six cases ; hemiplegic, four cases ; loss of power in left arm, ten cases ; in right arm, six cases ; in both arms and legs, one case ; in both arms, one case. The paralytic symptoms may precede, accompany, or follow the onset of the irregular movements. In the first group the diagnosis may present difficulties ; thus a girl was carried into my wards two years ago with the diagnosis of infantile spinal paralysis. In Case 89 (Infirmary Series) a girl, aged sixteen, with subacute rheumatism, had a sudden loss of power in the right arm and leg, with loss of voice, but without loss of consciousness. The attack lasted only half an hour, and was followed by pains in the limbs and choreic movements. The case was under observation for two months, and made a good recovery. In the majority of cases the weakness comes on during the attack, and is slight. The child may be unable to lift the arm to the head, the shoulder may droop, or the grasp may be feeble. Sometimes there is wrist-drop. Usually the weakness disappears with the cessation of the movements. Occasionally a local paralysis or weakness remains. In Case 229 (Infirmary Series) a lad of ten had severe general chorea in September 1880, with loss of power in the legs. He recovered, but when he returned in September 1883, in a second attack of chorea, he presented talipes of the left foot, the result of the paralysis in the former attack. In Case 21 (Infirmary Series) wrist-drop persisted for two years, the result of a palsy which came on with chorea of the right arm.

The monoplegic cases in young girls about the age of puberty may be difficult to separate from hysterical monoplegia until the choreic

<sup>1</sup> *Medical Gazette*. London, Vol. VIII, p. 849.



movements develop, or the diagnosis may be complicated by the existence of other conditions, as a tuberculous adenitis in a case recently reported by Massalongo.<sup>1</sup> There may be at first, too, difficulty in determining whether a patient has chorea minor or is the subject of post-paralytic movements. In Case 91 (Infirmary Series), a girl, aged five, had when two years old congestion of the brain followed by a hemiplegia, which disappeared in a few days. Since the attack there had been more or less general choreiform movements, which when she came under observation were slight but distinct, and under treatment disappeared in three months.

In severe cases of chorea with great jactitation the child may lose flesh rapidly and the muscles become thin and flabby. Occasionally in the affected limb the atrophy may be quite marked, and recently Raymond<sup>2</sup> has reported a case of marked monoplegic atrophy in the course of chorea. Perisson<sup>3</sup> has also written on the same subject.

<sup>1</sup> *Revue Neurologique*, 1893, Vol. I.

<sup>2</sup> *Société Médicale des Hôpitaux*, 1890.

<sup>3</sup> *Bordeaux Thesis*, 1890.



## CHAPTER III.

SYMPTOMS—(*continued*).

Disturbances of Sensation—Headache—Mental Symptoms—Cutaneous Symptoms—Urine, Nephritis—Fever—Duration—Recurrence—Chorea in Pregnancy—Chorea Minor in the aged.

**Disturbances of Sensation.**—Sensory troubles play a very much less important part in the history of chorea. Pain in the muscles or nerves of the affected limbs is not very common, but occasionally instances are met with, usually of hemi-chorea, in which pain without arthritic complication is a marked symptom. Mitchell has spoken of these as cases of "painful chorea." Some writers have laid special stress upon painful points over the sites of emergence of the spinal nerves, and it has been stated further that in instances of hemi-chorea the pain is only on the side of the spine belonging to the affected half of the body. The nerve trunks themselves may present special tenderness. Among French writers Triboulet and Marie have described carefully these sensitive points. I must say that in our experience at the Infirmary, and in my own personal experience subsequently, though I have frequently looked for these symptoms, they have been very rarely observed.

Numbness, tingling, and pricking sensations are occasionally met with. Hemi-anæsthesia, which has been noted, is usually an hysterical feature. Anæsthesia of the pharynx has also been observed (Comby).

Multiple neuritis occasionally occurs in chorea. Fry<sup>1</sup> has described the case of a girl aged 12, who had during a relapse of chorea increasing paresis, first of the lower extremities, and later also of the upper extremities, with atrophy and moderate contracture of the affected muscles, together with total anæsthesia, unpleasant sensations in the fingers and toes, and loss of reflexes. The muscles of the head, neck and trunk were uninvolved. The patient ultimately recovered. Railton<sup>2</sup> has reported a case in which the

<sup>1</sup> *Journal of Nervous and Mental Diseases*, 1890.

<sup>2</sup> *Medical Chronicle*, 1886.



paraplegia following chorea in a child aged 10 seemed due to neuritis.

Epileptiform seizures may occur, and in the Infirmary Series there are records of five cases with convulsive attacks or periods of sudden unconsciousness. Case 110 had a severe convulsion during the course of the disease. In Case 410, a child, aged 9, had had for five years epileptiform attacks, three or four in each year. When seen in May 1887, she was in a second, very severe attack of chorea. In Cases 202, 232, and 248 there were attacks of loss of consciousness, probably epileptic in character. Gowers<sup>1</sup> has called attention to the relation of chorea to convulsive seizures, and has reported ten cases.

Other forms of spasmodic disorders may coexist with chorea minor. Fry<sup>2</sup> has reported instances of athetoid movements, of rhythmical spasms, probably hysterical, and of tremor in cases of Sydenham's chorea.

The condition of the reflexes has not often been studied. Sinkler, in fifty cases at the Infirmary, found the knee-jerk normal in twenty-six, diminished in fifteen, while in nine it could not be obtained.

Trophic lesions are not common in chorea, unless we regard, as some would have us do, the joint troubles, as arthritis occurring in the course of a cerebro-spinal disease.

Headache is a frequent, and in some cases a persistent, symptom. It may precede an attack for some days. It may be paroxysmal. The special senses are rarely involved. The question of eye-strain has already been fully considered. Among rare eye symptoms may be mentioned embolism of the retinal artery, of which cases have been reported by Benson,<sup>3</sup> Swanzy,<sup>4</sup> Sym<sup>5</sup> and Ball.<sup>6</sup> Minute retinal hæmorrhages may also be due to embolism. Gowers states that in a few cases optic neuritis occurs, and he speaks of one case in which it was comparable to that seen in brain tumour. Gordon tells me that he has observed retinal hyperæmia in many cases.

**Mental Symptoms.**—Psychical disturbance is rarely absent in chorea; fortunately in the majority of the cases it is slight in degree. The following are the most striking of the mental changes in the

<sup>1</sup> *British Medical Journal*, 1878.

<sup>2</sup> *Journal of Nervous and Mental Diseases*, 1892.

<sup>3</sup> Referred to in *Lancet*, January 30, 1886, p. 219.

<sup>4</sup> *Ibid.*

<sup>5</sup> Quoted by Gowers.

<sup>6</sup> *Clinical Society's Transactions*, Vol. XXI.



disease: (1.) Disturbances of the moral sense, manifested frequently in a strange perverseness, great irritability of temper, with emotional outbreaks. A frequent complaint heard from the mother is that the character of her child is completely changed. A patient may do odd and meaningless acts, thus, Case 150, a girl of 21, in a second attack had the trick of hiding away her clothes, and was in the habit of wandering off by herself. On one occasion she went off and was not found for two days.

(2.) Disturbances in the faculties of memory and attention. The aptitude for study is lost, and the child may no longer take an interest in story books. This is of course very much more marked in some cases than in others. Transient loss of the power to read and to write may exist. In the extreme instances we can often "read the mind's complexion in the face," and can see, as the disease progresses, a cloudiness obscuring the bright, clear countenance, leaving it dull, heavy, and expressionless. Actual melancholia may occur; in rare instances the impairment of intellect may be progressive and terminate in dementia. Marcé,<sup>1</sup> whose essay on the subject remains the most important, and Axenfeld and Huchard<sup>2</sup> lay great stress on hallucinations by day and in the intervals between sleeping and waking.

(3.) While the great majority of the cases of chorea minor display nothing more than slight mental changes, the rarer, more aggravated forms, coming under the head of chorea insaniens, present, as their most striking characteristics, hallucinations, delusions, and mania. So striking may these features be, that there are many instances on record in which the bodily trouble has been entirely overlooked, and the patient has been committed to an asylum.

Whether there is a chronic form of insanity deserving the name of *folie choreique* is extremely doubtful.

**Cutaneous Symptoms.**—Skin affections in chorea are not very uncommon. Some of them are due no doubt to the disease itself; others follow the prolonged administration of arsenic, which is so commonly employed in this disease. Out of 410 of the Infirmary cases there were eleven with skin affections. In five of these the rash was erythematous or papular, and unquestionably due to arsenic. There were two cases of herpes zoster. In Case 2, a girl of 7, the rash came out on the right side about three weeks after she began

<sup>1</sup> *De l'état mental dans la Chorée. Mémoires de l'Académie Impériale de Médecine*, XXIV.

<sup>2</sup> *Traité de Névroses*. Second Edition, 1883.



to take arsenic. In Case 318, a girl of 11, the herpes was on the left side in the lumbar region, and appeared seven weeks after the beginning of the use of arsenic. Pigmentary changes in the skin have been frequently noted in chorea, and are probably always a consequence of the prolonged administration of Fowler's solution. In Case 332, a boy aged 7, there were pigmentary changes on the right cheek after a six weeks' course of Fowler's solution.

Gradual pigmentation from the excessive use of arsenic is quite well recognized, and many such cases are to be found in the literature.

Among the most interesting cutaneous manifestations of chorea are those associated with arthritis, which are very similar to those developing in so-called rheumatic purpura. The most common are those which take the form of multiple erythema, either an erythema nodosum, a purpuric urticaria, or sometimes a simple purpura. In Case 13, a girl aged 18, without any rheumatic manifestations, had an eruption resembling that of erythema nodosum, which persisted for some days. In Case 312, a girl aged 8, with pains in the wrists, at the time of the attack black and blue raised spots appeared on the legs; they came out in crops and were not painful. They showed themselves before the administration of arsenic. In Case 347, a child aged 8, had a first attack of chorea in 1885 and a second in 1886, the latter having been preceded by an attack of acute rheumatism. The knees and ankles were swollen, and there were large purple blotches on the skin of the legs and of the arms.

In four of the recent fatal cases (see Morbid Anatomy) there were skin rashes in two resembling the scarlatinal eruption of septicæmia, in one purpura, and in one there was the macular rash of secondary syphilis.

**Urine in Chorea.**—An increase in the amount of urea was determined by Walshe, Bence Jones, Kelly and others. Handfield Jones found the phosphates increased. Albuminuria is not uncommon in the very severe forms, and glycosuria has been described. Herter (private communication) found the uric acid much increased, and in several cases the chlorides were diminished. Todd, in his "Lectures on Diseases of the Nervous System," says that the uric acid may form a sediment, as in gouty conditions.

A. E. Garrod<sup>1</sup> found uro-hæmatoporphyrin present in the urine of fourteen out of twenty choreic cases. This substance is described

<sup>1</sup> *Lancet*, 1892, I.



by McMunn as particularly frequent in the urine of rheumatic persons, and Garrod looks upon this as an evidence of the close relationship of the two diseases.

Nephritis is probably not so rare as has been thought. Thomas,<sup>1</sup> of Freiburg, has reported a case of a boy of 14, who had not previously had either chorea or nephritis, and who, three weeks after the first outbreak of chorea, had general anasarca and the signs of acute nephritis. Prof. Thomas thinks there was an etiological connection between the two.

In Case 6 of the Guy's Hospital series reported by Goodall,<sup>2</sup> a girl aged 17, who had never had rheumatism, was admitted in her second attack of chorea. Pericarditis was detected. She died suddenly and unexpectedly. There was slight pericarditis, and recent endocarditis of aortic and mitral valves. The kidneys weighed 13 ounces, were enlarged, red, with swollen cortices, and showed all the signs of an acute nephritis.

In one of Turner's cases (No. 60 of series of fatal cases), a girl of 17 had acute nephritis for a week, with hæmaturia, and No. 12, a girl of 11 years, also had hæmaturia.

**Fever in Chorea.**—Except in the severe maniacal form, fever is rare. In the Collective Investigation Report of the British Medical Association pyrexia was stated as being present in only 12 per cent. The study of the disease in out-patients does not favour thermometric investigation. Of twenty-eight cases treated in my wards at the Johns Hopkins Hospital there was only one, a case of chorea insaniens, in which the temperature rose to 105.2°. In eleven instances the temperature registered above 100°; in six of these above 101°. As a rule, anything more than a slight rise of temperature is usually to be referred to a complication; thus, Jos. C., Medical No. 3,064, had been ill for two weeks with pain and swelling of the feet, and chorea. When admitted, July 3rd, 1893, he had a temperature of 104°. The choreic movements were active. There was a soft systolic murmur. Under the salicylates the temperature fell, was normal on the 6th, and remained so. In other cases the fever seems due to an endocarditis, or to an endo-pericarditis. Occasionally one meets with cases in which it persists for days without any obvious cause except the chorea itself; thus, in the case of Marion S., admitted July 11th, 1892, Medical No. 2,191, the patient had had chorea for several weeks without any articular

<sup>1</sup> *Neurologisches Centralblatt*, Bd. XI., p. 391.

<sup>2</sup> *Guy's Hospital Reports*, Vol. XLVII.



affection and without any heart symptoms. The temperature from the 11th to the 18th ranged from  $99.5^{\circ}$  to nearly  $101^{\circ}$ . A still more striking case is the following:—Laura D., aged 14, Medical No. 1,527, admitted September 3rd, 1891, in her third attack of chorea, which had lasted for several weeks. She had not had articular trouble, but her father has had rheumatism. On admission she had chorea of moderate severity; the heart was not affected. The temperature was  $99.3^{\circ}$ ; on the 6th and 10th it rose to  $100^{\circ}$ . She complained frequently of being chilly. On the 12th the temperature rose to  $103^{\circ}$ , and in the evening nearly to  $105^{\circ}$ . There was no joint affection, and the throat was not sore. The temperature kept up, and for the next three days was frequently in the neighbourhood of  $104^{\circ}$ . Then from the 15th to the 18th it ranged from  $100^{\circ}$  to  $102^{\circ}$ . On the 19th it became normal. We were at a loss to know exactly upon what this fever depended. There seemed to be slight enlargement of the spleen; the choreic movements persisted, but there was no sore throat and no heart complication. It was suggested by Dr. Thayer that possibly this might be an illustration of Kahler's non-articular rheumatic fever,<sup>1</sup> and on the 18th she was ordered sodium salicylate, five grains every two hours. The temperature at the time was  $102^{\circ}$ . At 4 A.M. on the 19th it was  $98.7^{\circ}$ , the lowest point reached since entrance. At 6 P.M. on the 19th it was  $101^{\circ}$ , and fell to  $99^{\circ}$  in the evening. The salicylates were kept up for three days, and she had no more fever until November 8th and 9th, when she had a rise of temperature to nearly  $103^{\circ}$ . She was discharged well on November 23rd.

Finlayson<sup>2</sup> (Glasgow) has written an interesting paper upon short febrile attacks in chorea, which he suggests are due to the endocarditis. He gives the case of a child aged 7, admitted to hospital November 18th, 1884, with a chorea of the left side of seven weeks' duration. There was no history of rheumatism. There was an apex systolic murmur. The chorea improved, but on December 14th the child began to have fever, the temperature rising to  $103^{\circ}$  without any local manifestations. There was variable fever throughout January, in February it increased, and the range was from  $102^{\circ}$  in the morning to  $105^{\circ}$  in the evening. Post-mortem there was found acute endocarditis of the mitral and tricuspid segments, with very slight fresh pericarditis. It is important to bear in mind that the fever may also be kept up by pericarditis, as in the case reported

<sup>1</sup> *Zeitschrift f. Klin. Medicin*, Bd. XIX.

<sup>2</sup> *Archives of Pediatrics*, Vol. VII. p. 97.



by Sinkler,<sup>1</sup> of which I give an abstract in the section on the heart.

The temperature may be increased on the affected side. H. A. Hare<sup>2</sup> has reported several such cases; one of his patients said that he felt the hand getting hot, as the movements became exaggerated in the limb.

In cases of maniacal chorea, and in the severer forms without delirium, fever is usually present, and may reach a high grade. In one of Mitchinson's cases (No. 18) the temperature was 107.6°. In a case reported by Grosse (No. 16) it was 41.5° C., and in No. 11 (Donkin and Hebb) the thermometer registered 107°. There may be, however, the most severe chorea with complications, pneumonia and pericarditis, without any fever to speak of, as in the case of Neuwerck (No. 66).

**Duration.**—Very mild cases may terminate in three weeks, but from eight to ten weeks may be stated to be the average duration of attacks of moderate severity. Cases, however, very frequently drag on for three or four months, and there are instances of true chorea minor in which the movements persist for even six months. The term acute chorea may very properly be applied to the very severe cases with fever and delirium, which may terminate within a week or two after the onset. The very chronic cases in children and in young adults are very rarely true chorea minor, but belong to a different category, and are usually forms of tic. The instances of congenital chorea and chorea spastica have nothing to do with the form at present under consideration. The following cases, however, may be instances of unusually protracted chorea minor:—

Case 186, Infirmary Series, Lizzie K., aged 21, no history of rheumatism. When thirteen years old she had the first attack, which was general and affected the speech. After lasting without intermission for three years the movements ceased, and she returned, December 1st, 1882, with a second attack, which had already lasted three months.

Case 335, Infirmary Series, Mary M., aged 12. Mother very nervous, and an uncle had spasms. For three years she had had movements in the face, head and arms. The speech was never involved. On September 21st, 1885, when first seen, the head was chiefly affected. Jerking movements of the hand also occurred when at rest. She was treated for some months without any im-

<sup>1</sup> *University Medical Magazine*, 1890, p. 483.

<sup>2</sup> *Boston Medical and Surgical Journal*, Vol. CXII.



provement. On the 7th of May, 1887, she returned to the clinic. The choreic movements had gradually disappeared, and though nervous she seemed quite well.

The following is a very remarkable case, which was under observation for some years:—

Alfonso G., aged 21, baker. His sister had chorea, and died of heart disease in 1872. In August, 1884, he began to have jerking movements in the head and arms and legs. The speech was also somewhat affected. When seen June 1st, 1885, there was spasmodic contraction of all the muscles of the face, neck, trunk and arms. The legs were not affected. The inspiratory muscles were involved, and at times there was high-pitched whistling inspiration. The heart was not affected. He remained under observation and treatment for nearly three years, during which time the movements persisted with very little variation in intensity. When I last saw him the twitching and jerking of the muscles of the neck and chest were present, and the case then looked more like an instance of habit spasm.

It is quite possible that the localized movements of a true chorea minor may persist as facial tic, and Leroux<sup>1</sup> has reported cases of this post-choreic tic, but they are of extreme rarity.

**Recurrence.**—The liability of chorea to recur was noticed by Sydenham, who says that “as the disease is liable to return again, I think it well for the patient to be purged about the same time, or a little earlier, the following year.” In the 410 cases analyzed with reference to this point, in 240 there was a single attack; in 110, two; in 35, three; in 10, four; in 12, five; and in 3, six. Case 23 of the Infirmary Series is a good illustration. A girl, aged 8, came to the clinic for the first time March 23rd, 1877. The attack had begun in the autumn of the previous year. She improved under treatment, and the movements ceased. Towards the end of June the general symptoms returned, and this is noted as the second attack, though it probably was only a relapse. The third attack was in August, 1880, the fourth in the spring of 1881, the fifth in January, 1882, and in February, 1883, she returned with a sixth attack. There was no rheumatism, and the heart was not affected. Gowers mentions a case in which there were nine attacks.

We know very little as to the cause of the recurrences. Females appear to be more liable to them than males, but this is only natural. It is doubtful whether rheumatism has any influence. In cases with

<sup>1</sup> *Revue des Maladies de l'Enfance*, 1891.



frequent recurrence heart disease is more common ; thus, of the seventy-two cases to be hereafter noted with organic disease following chorea, thirty had had three or more attacks of chorea. Mitchell has called special attention to the vernal recurrence of chorea, and at every clinic there are many instances in which during the months of March, April, and May, children are brought back with fresh attacks of the disease. It was a custom at the Infirmary for Diseases of the Nervous System to warn the parents on this point, and give instructions to be particularly careful, and even to resume the arsenic in the spring. The recurrence may, however, be autumnal.

The interval between the attacks is usually many months, or it may be several years. In a majority of the cases as long a period as twelve months intervenes. Fright and over-work at school are often mentioned as exciting causes of the recurrence. The succeeding attack may be much more severe than the first. The same muscles are usually involved in the recurrence as were affected in the primary attack, but this is not always the case. Other muscles may be involved, and though the recurrence is apt to be of less intensity, there are instances in which it is very much more severe. The heart may not be affected until after the third attack, and as in the case above mentioned, even a series of attacks may occur throughout several years without any cardiac involvement.

#### CHOREA AND PREGNANCY.

The association was noticed by several observers during the last century. Important statistical papers have been written by Barnes, Mosler, and others.

The following cases have occurred at the Infirmary :—

*Case 277.*—A woman, aged 20, who had had repeated attacks of chorea, applied at the clinic October 15th, 1884. She was married early in May. Began to have movements of limbs August 15th. At time of visit both sides were involved, and there was decided affection of speech. She had morning sickness, the menses had stopped, the breasts were enlarging, the papillæ prominent, and she was believed to be pregnant. She attended at the clinic for four weeks, and improved very much. The mental condition was good. There is no note as to her complete recovery.

*Case 236.*—Mrs. B., aged 22, applied at the clinic November 16th, 1883. She had an attack of chorea when nine years of age, and has been nervous ever since. Is pregnant. Attack began about



September 15th, and has recently got worse. The right side chiefly involved; speech much affected; no mental symptoms. She remained under treatment for a few weeks, improved slightly, but did not return.

A third case was admitted to the Hospital in the spring of 1876. The notes have been mislaid, and I am indebted to Dr. Neff for the following memoranda. She was aged about 30, a servant at the Colonnade Hotel. She had had chorea as a child. In the fourth or fifth month of pregnancy she accidentally broke a demijohn of brandy, and received a severe shock. Soon after chorea developed, and she was brought to the hospital. The attack was very severe, and after two months she was transferred to the maternity department of the Philadelphia Hospital, where she died.

A fourth case was under treatment by Dr. Sinkler in 1887. Woman aged 21, married three months. Menses had stopped and she was believed to be pregnant. Nine years ago she had chorea—a most obstinate attack—which persisted for five years without intermission. She had a second attack, which lasted two months. The present one began shortly after her marriage, and is, possibly, not directly dependent upon pregnancy. It is, so far, not a serious attack.

The following case came under my observation at the University Hospital:—

Jennie C., aged 18, seen on the 4th of June in the Medical Dispensary of the University Hospital.

She was married last March. Healthy and well as a child, and as a young girl; has not menstruated since marriage; believes that she is pregnant. A week ago she noticed jerky movements of the left arm, the left leg, and the left side of her face: had been feeling nervous sometime before. She is somewhat anæmic.

She now has distinct hemi-chorea with irregular movements of the hand and arm, but less marked movements of the foot and leg; the left side of the face twitches; when the tongue is protruded it moves about irregularly; she complains of loss of power in the left arm; pulse 96; temperature not elevated.

The apex beat of the heart is in normal position; there is soft systolic murmur obliterating the first sound; it is not heard at mid-axilla. In the fifth interspace toward the sternum both sounds are heard with distinctness; in the second, third, and fourth left interspaces there is a loud systolic murmur with marked accentuation of the second sound; no murmur at the aortic cartilage.



Subsequently this case came under the care of Dr. Wood and Dr. Hirst, the latter of whom has reported it.<sup>1</sup> The history is of interest, as it appears that her unilateral chorea improved very much but she became profoundly apathetic and melancholy, and had very pronounced anæmia. The choreic movements stopped entirely. So serious, however, was her mental condition that it was thought advisable to bring on premature delivery, which was done successfully. The prolonged apathy and melancholy is seen also in children. In a case which was in my wards last winter the condition persisted for more than three months.

Primiparæ are most frequently attacked, and a previous chorea plays an important rôle ; thus out of thirty-four recent cases collected by McCann,<sup>2</sup> eleven had had chorea previously. In his statistics he does not give the number in which it occurred during the first pregnancy. Fright, mental emotion, and anæmia are mentioned as special factors.

In his paper he gives the following statistical details:—In thirty-seven cases the ages were as follows:—At seventeen, 3 ; at eighteen, 4 ; at nineteen, 6 ; at twenty, 11 ; at twenty-one, 2 ; at twenty-two, 3 ; at twenty-three, 3 ; at twenty-four, 1 ; at twenty-five, 1 ; at twenty-six, 3.

The period of occurrence during pregnancy was as follows:—“ Out of 36 cases, first month, 2 ; second month, 6 ; two-and-a-half months, 1 ; third month, 6 ; three-and-a-half months, 2 ; fourth month, 7 ; fifth month, 4 ; five-and-a-half months, 2 ; sixth month, 3 ; six-and-a-half months, 1 ; seventh month, 1 ; eighth month, 0 ; ninth month, 1.”

The following details as to causation are given:—“ Out of 34 cases, chorea previously, 11 ; rheumatic and scarlet fevers, 2 ; rheumatic fever alone, 2 ; fright only cause stated, 2 ; no cause stated, 4 (in one of the cases one sister had had rheumatic fever ; in another chorea and rheumatism existed in the family) ; rheumatic fever and fright, 1 ; rheumatic fever and chorea, 7 ; scarlet fever, rheumatic fever, and chorea, 1 ; chorea and fright, 3 ; mental distress owing to pregnancy, 1.”

Out of 32 cases in which there was no artificial interference the patient was delivered at term in 26. In one case there was accidental hæmorrhage and miscarriage at the fourth month ; in one case miscarriage at the fifth month ; and in four cases miscarriage at

<sup>1</sup> *University Medical Magazine*, Philadelphia, Vol. I.

<sup>2</sup> *Transactions of the Obstetrical Society of London*, Vol. XXXIII.



the sixth month. Out of 39 cases death occurred in 7 ; in 3 from mania ; in 3 from exhaustion ; and in 1 from puerperal peritonitis.

In the 72 fatal cases which I have collected from the literature since 1881 only four cases were associated with pregnancy. This is a very much smaller percentage than in Raymond's tables, in which of 79 fatal cases 11 were in pregnant women.

Post-partum chorea is much more rare. McCann refers to only three or four cases, but he states that it is not yet definitely settled if this form of the disease exists. The following instance, reported by Litten,<sup>1</sup> seems to be genuine :—

A primipara was delivered on December 3rd of a living child. On the sixth day after delivery she was sent to the medical clinic with an extensive scarlatinal rash and high fever. On the third day after admission the rash faded, but the fever kept up. On the evening of December 9th she had a chill, and many of the joints became swollen and painful. The lochia were offensive. On the 13th of December choreic movements began in muscles of limbs, which became general and severe. There was irregular fever, which with the arthritis made up a picture of puerperal fever. Death on December 19th. Autopsy : Diphtheritic endometritis ; polyarthritis ; purulent septic infarcts ; heart normal.

It is to be borne in mind that hysterical disorders of motion may occur post-partum.

#### CHOREA MINOR IN THE ADULT AND IN THE AGED.

The comparative rarity of the disease in the adult is well shown in the statistical table, which gives in the third decade only ten cases ; in the fourth decade only one ; and above the fourth only two. Sinkler<sup>2</sup> has reported two cases in patients over eighty. Saundby<sup>3</sup> has collected twelve cases. Many of the cases reported as chorea belong in reality to affections quite different from chorea minor.

Herringham,<sup>4</sup> who has given a critical analysis of the cases described as chorea in the aged, divides them into four groups : first, chorea minor ; second, choreiform movements due to coarse sub-cortical lesions ; third, the chronic progressive chorea of Huntington ; and fourth, a chronic chorea of adults and the aged, without hereditary bias as in the Huntington's form, but with coarse cortical changes, and accompanied by mania and dementia. The

<sup>1</sup> *Charité Annalen*, Bd. XI. p. 279.

<sup>2</sup> *Journal of Nervous and Mental Diseases*, July, 1881.

<sup>3</sup> *Lancet*, 1884, II.

<sup>4</sup> *Brain*, Vol. XI, 1889.



following is a good illustrative case of Sydenham's chorea in an old woman :—

Lousia B., mulatto, aged about seventy, was admitted December 2nd, 1893, with choreic movements in the hands and arms, particularly on the left side. The patient was a widow, and had had nine children, all of whom are dead. She was a domestic in the family of a lady who had promised to remember her in her will, but, failing to do so, the patient was very much worried and distressed, and about four or five weeks before entrance she began to have twitchings, and it was stated by the friend who brought her that she had not been quite right in her head. It was impossible to get a satisfactory history as to the date of the onset, but she was positive that she had never had any similar trouble before, and of the association of the attack with disappointment and the loss of her mistress.

On admission her temperature was normal ; she was somewhat irrational ; could not be kept quiet, and would not stay in bed. She talked in an irrational manner, though she evidently understood what was said to her. She walked fairly well ; the arms and hands displayed quick movements, and there were occasional jerking of the muscles of the face. The muscular power was good. There was a loud, blowing systolic murmur heard over the whole cardiac area, but the heart did not appear to be hypertrophied. The urine was normal. The patient was ordered to be kept in bed and have Fowler's solution of arsenic, three minims three times a day.

Throughout December she improved a good deal and the movements were very much less, being confined chiefly to the left arm and hand, and the left side of the face. Early in January they increased somewhat. She was able, though, to be up and about, and her mental condition seemed somewhat better, though she was rather stupid and dull.

Throughout January and February she improved a good deal, but the movements never entirely ceased. She was able to be up and about the ward.

In March the condition was unchanged. The movements were very slight, being confined chiefly to the left side of the face, and to the left hand and arm. Sometimes they were scarcely noticeable. Early in April she had one or two attacks of cardiac dyspnoea and had cough. On the evening of the 7th she had a very severe attack, in which she sat up, gasped for breath, became quite pulseless, and died in a few hours.



*Autopsy.*—There were no coarse cerebral lesions. The heart was hypertrophied, the mitral segments were thickened and insufficient from old sclerotic endocarditis; there were no fresh vegetations.

Herringham<sup>1</sup> gives the following abstracts of chorea minor in the aged and adult, which I here add: "Roger has recorded a case in which an old lady of 83 was affected by an ordinary chorea which came on without cause, and was cured in five weeks; Russell writes of a woman of 77, who had it mainly on the left side, and recovered in three months; Sinkler gives a case of a man of 86 who recovered in a few months; Saundby gives two cases, but the histories do not extend for a sufficient length of time to enable them to be classified. Ferguson had a case of a woman of 74, who after much fatigue in nursing her husband, became somewhat irritable and excitable, and then began to twitch on the left side. She recovered in eleven weeks. In the Collective Investigation Report, there are several cases which Dr. Mackenzie has grouped together as chorea at exceptional ages. No. 119, by Gowers, is a second attack of chorea following rheumatic fever in a man of 40. No. 223, by Alexander, is a woman of 63, in whom the attack lasted four months. She also had had it before. No. 222, by the same author, is a first attack in a woman of 68, lasting six months. Carline records a case, No. 180, of an old woman of 73, in whom a chorea lasted four weeks. She had had several attacks of late years. Aitken describes another case, No. 93, of a woman of 86, lasting one year. Of this kind also is Graves's case of a man of 70, in whom the attack lasted several months, and Gauthier records an attack in a woman of 75, due to peripheral irritation from extraction of a tooth, similar to those occasionally met with in the young. She recovered after fourteen days."

The cases must be distinguished carefully from generalized forms of tic, and from the disordered movements associated with organic lesions. There is less likelihood of confusion with Huntington's chorea. The disease may prove of great severity, and run a malignant course. One of Koch's cases (No. 50 in the table) had had several previous attacks. Fresh endocarditis was found. No. 17, a man aged 66, had had chorea eight years before, and died in coma on the fourth day after admission, possibly from the effects of bromide of potassium and chloral. Fresh vegetations were present on the mitral segments. In No. 71 the chorea followed emotional disturbance in an old woman with mitral insufficiency. Death occurred in an attack of cardiac dyspnoea. There was no fresh endocarditis.

<sup>1</sup> *Loc. cit.*



## CHAPTER IV.

## THE HEART IN CHOREA MINOR.

Condition of the Heart during the Attack—Condition of the Heart in Fatal Cases  
—Subsequent Heart History in Choreic Patients—Pericarditis in Chorea.

RARELY a fatal disease, a series of several hundred cases of chorea may be treated without a death ; nor does it often leave permanent damage of the nervous system, since, so far as intelligence and motor power are concerned, the children may recover perfectly. The only serious event is the occurrence of endocarditis, and the danger here, not immediate but remote, lies in the changes which may be initiated by the acute valvulitis. A study of the cardiac relations of chorea must embrace (1) the condition of the heart during the attack ; (2) its condition in fatal cases ; (3) the subsequent heart history of persons who have recovered from the disease ; and (4) the state of the pericardium.

## I.—CONDITION OF THE HEART DURING THE ATTACK.

The bare chest should be examined. Auscultation through the clothing is not trustworthy, since soft murmurs, readily audible with the stethoscope placed directly against the chest wall, may escape detection. It is a good plan to let the child remain quietly on a lounge for some time, and make the examination first in the recumbent posture. Subsequently the effect of exercise and of the erect position may be tested. In the severer forms of chorea the extreme jactitation may prevent for some days a satisfactory examination. Children with chorea rarely complain of palpitation, pain about the heart, or of symptoms which would direct attention to the organ. As in acute rheumatism, it is for us to look for evidences of disturbed action.

Anomalies in the cardiac action may depend upon faulty innervation, upon changes in the muscular walls, or in the condition of the circulating blood, and upon inflammation of the valve segments.

Palpitation and disturbance of rhythm, rapid action, and pain are



the chief nervous manifestations. Palpitation and irregularity are not very common, and are not so frequent as the rapid action under emotional disturbance. Occasionally the heart-sounds have a foetal rhythm. Cases with very exaggerated movements may present for days an excessively rapid heart action. On the other hand, with the mental enfeeblement which sometimes follows chorea, the pulse may be abnormally slow, beating in a child of ten or twelve at the rate of seventy or eighty a minute. I have never seen a case in which the disordered movement was of such a kind that it might be attributed to a special choreic action of the heart muscle (*chorée du cœur*).

The subjects of chorea rarely complain of pain about the heart, and even with very rapid action and palpitation there may be no subjective sensations. Pain as a marked symptom is met with in recurring attacks associated with endo-pericarditis and rheumatism, more rarely in the first attack with endocarditis.

Evidence of involvement of the myocardium, of the valves, and of disturbed function due to impoverished blood is afforded by the presence at one or other of the so-called cardiac regions of the abnormal sound known as a murmur, the nature of which in chorea has been so much discussed.

It may be well to speak first of the incidence of heart murmurs during the attack. Of the 554 cases at the Infirmary for Diseases of the Nervous System, 170, 30·7 per cent., presented heart murmurs; in 149 apical in maximum intensity; in 21 basic. In 141 cases of chorea minor examined at the Dispensary for Nervous Diseases of the Johns Hopkins Hospital, there were 42 with a cardiac murmur. Of the 449 cases in the Report of Committee on Collective Investigation of the British Medical Association, 113 had heart murmurs.

Much has been written in explanation of the occurrence of heart murmurs in this disease, and an excellent summary is to be found in Hayden's<sup>1</sup> work.

The murmurs may be either functional or organic.

*Functional.*—The basic systolic murmur heard usually with greatest intensity in the area of the pulmonary artery, but audible sometimes in the aortic area, may be due to the excited and very rapid action of the heart. A murmur of similar character is common in neurasthenic women, and may be heard in thin-chested subjects after violent exercise, and in children with the rapidly acting heart of high fever. I have had many opportunities of noting, after rest

<sup>1</sup> *Diseases of the Heart and Aorta*, 1875, Part I, pages 265-277.



in bed for a day or two, the disappearance of a systolic murmur at the base, which had been heard with great clearness during the excitement of the first examination of the child in the Dispensary.

With anæmia and debility, frequent associates of a chorea in its third and fourth weeks, there may develop soft systolic murmurs in the pulmonary artery and apex areas, often, too, heard intensely over the body of the heart along the left sternal margin. Frequently with it one notices a wide area and fulness of cardiac impulse, and sometimes systolic pulsation in the cervical veins. The murmurs may be audible only in the recumbent position, disappearing when the patient stands up. These murmurs are in all probability caused at the pulmonary and tricuspid orifices, and are accompaniments of debility and anæmia.<sup>1</sup> In protracted cases with marked debility, and weakness of the heart muscle, the systolic apex murmur may be mitral in origin and be due to muscular insufficiency.

Another explanation of the frequency of heart murmurs in chorea is the theory "that irregular and occasional reflux takes place at the mitral orifice through disordered action of the muscular apparatus connected with the valve" (Walshe). The objection to this view which Kirkes urged years ago, that there was no proof of the participation of involuntary muscular organs in the choreic disorder, still holds, nor is there such inconstancy and variability in the apex heart murmurs of chorea as would be inevitable did the condition result from valvular insufficiency in consequence of a "want of correspondence between the fibres of the ventricle which obliterate the cavity and those that close the valve."

There is experimental evidence to show that one set of the papillary muscles may be completely functionless without causing any murmur. Dr. Townsend Porter, of the Harvard Physiological Laboratory, has shown that ligation of the ramus descendens of the left coronary artery causes within twenty-four hours complete infarction of the anterior papillary muscles from which the chordæ tendineæ pass to one-half of each mitral flap. On auscultation at a time when infarction was fully developed no murmur was noted on repeated auscultation in several animals.

In a large proportion of all cases of chorea in which a murmur is heard at the base, or along the left margin of the sternum in the second, third and fourth interspaces, the disturbance is probably functional.

<sup>1</sup> See discussion of their causation in *Investigations into some Morbid Cardiac Conditions*. William Russell, M.D., Edinburgh, 1886.



*Murmurs of Organic Origin.* — Acute endocarditis, commonly of the mitral leaflets, occurs with great frequency in chorea, and the remarkable statement that there is no other disease, not even acute rheumatism, which is so frequently accompanied with valvulitis, seems quite justifiable. The apex systolic murmur heard in many cases of chorea is doubtless due to the endocarditis. The symptoms and physical signs of acute endocarditis are very uncertain; the mitral segments may be inflamed, and yet the patient may present no cardiac symptoms whatever. There are very carefully observed cases of chorea, in which the apex murmur has not been present, and yet post-mortem the mitral segments have shown vegetations. Feelings of oppression about the heart, palpitation, transient dyspnoea, though mentioned as symptoms of acute endocarditis, are, after all, rarely present in the first attack, and are much more commonly seen in a recurring endocarditis attacking a heart already damaged. Fever, too, is a variable symptom, and is not always present.

The physical signs, though in a more reliable, are nevertheless inconstant. The apex beat may be a little diffuse and the area of dulness increased, both phenomena being due to slight dilatation of the left ventricle. An alteration of the first sound, which has a prolonged or dull character, with the subsequent occurrence of a blowing murmur at the apex region, developing under observation in a case of chorea, rheumatism or fever, and more particularly in the earlier stages, before the patient has become much enfeebled and anæmic, are the most reliable auscultatory signs.

The following statements may be made on this question :—

1. The extraordinary frequency with which mitral valvulitis is met with in fatal cases is remarkable. *There is no known disease in which endocarditis is so constantly found, post-mortem, as chorea*; it is exceptional to find the heart healthy.

2. The character and location of the apex murmur are such as experience in other affections has taught us to be associated with inflammation of the mitral segments. Why this murmur should be so generally connected with the presence of a row of small warty vegetations just within the auricular margins of the curtains, and not capable, as one would think, of seriously interfering with their functions, is a problem to be solved. The condition certainly does not necessitate regurgitation, and the bruit may perhaps, as has been suggested, be due to friction of the roughened faces of the segments.



3. The inconstancy of the murmur and its disappearance on the subsidence of the chorea have been urged against this view. As already stated the bruit may be variable, and, indeed, does not necessarily accompany mitral endocarditis. Kirkes, years ago, insisted upon this, and there have been two autopsies in carefully studied cases of chorea in which the vegetations were found post-mortem, although careful examination during life had failed to reveal a murmur (Baxter: *Brain*, Vol. II.; Frank. *Allg. Wiener Med. Zeitung*, 1879). There are facts which suggest that we may during the attack have an endocarditis, not manifested even by a murmur, which has nevertheless laid the foundation of future trouble. The disappearance of the apex murmur of chorea—and of rheumatism also—has been repeatedly followed; and if it is true that the murmur is caused by the small vegetations, that it should disappear is only a natural sequence of the changes which go on in them. At first a soft granulation tissue, they become in time firmer and smaller, until ultimately smooth flat elevations mark the spots. It is not improbable that if we could follow accurately the auscultatory history of a valve affected with acute endocarditis, we should find in many cases that the murmur of the fresh attack disappeared, to reappear when the changes, which it is the misfortune of the acute disease to initiate, have reached a point at which they begin to interfere with the competency of the valve.

4. In its sequel the cardiac affection of chorea has been supposed to differ from that of other diseases, "as none of the injurious after-consequences which attend endocarditis in its other relations, . . . are found to ensue here" (Sturges). The examination of a large number of choreics some years subsequent to the attack tells a sad tale to the contrary, and proves that the primary heart trouble is, in a majority of cases, at least, an endocarditis.

## II.—CONDITION OF THE HEART IN FATAL CASES.

The statistics of fatal cases of chorea have been collected by Sturges<sup>1</sup> and Raymond.<sup>2</sup> Sturges states that of eighty cases representing the combined experience of Guy's, St. Bartholomew's, St. George's, and St. Thomas's Hospitals, there were only five in which the heart valves and pericardium were reported healthy. In Raymond's table of seventy-nine cases, if we exclude the London cases,

<sup>1</sup> *Chorea*. London, 1881.

<sup>2</sup> *Dictionnaire Encyclopédique des Sciences Médicales*. I Série, 25. Art. Danse de Saint Guy.



thirty-four are left; in only nineteen of these are specific statements given as to the condition of the heart, and in every one of the nineteen endocarditis was present. I have collected from recent literature seventy-three additional cases, of which sixty-two presented endocarditis. The table of these cases will be found under an appendix to the section on "Morbid Anatomy." My personal experience includes five cases, in four of which endocarditis was present. Brief notes of the cases are as follows:—

*Case I.*—S., girl, aged 11, had had acute rheumatism. Admitted to the Montreal General Hospital, under Dr. George Ross, with acute chorea, and died of an intercurrent pneumonia. The movements had almost ceased under hypodermics of arsenic. The autopsy showed slight hypertrophy of the heart, somewhat thickened mitral curtains, with numerous, irregular, warty vegetations just inside the auricular margins. Two of the aortic segments also presented bead-like vegetations below the corpora Arantii.

*Case II.*—T. B., a boy, aged 11; had chorea in May, 1880, and a second severe attack in July of the same year. No rheumatism. No heart murmur. About the 20th of February, 1881, there was a recurrence, and on March 3rd he came to the General Hospital to see Dr. Molson. About the 10th he began to get feverish and extremely restless. On the 14th the temperature was above 104°, and he became comatose. The left arm seemed powerless; the right arm and leg were constantly twitching. On the 15th the temperature reached 105° F., and there were cutaneous ecchymoses. He died on the morning of the 16th. The autopsy showed very extensive mitral valvulitis, the vegetations large, soft, greyish-white in colour. No chronic affection of the valves. The spleen and kidney contained many recent infarcts. The brain and membranes healthy, with the exception of a spot of greyish-red softening in the right corpus striatum (lenticular nucleus) about the size of a cherry. It was no doubt embolic, though the arteries of the perforated space were carefully examined for emboli without success.

*Case III.*—Emma M., aged 18, was admitted to the Montreal General Hospital, under Dr. George Ross, and died in five days of exhaustion. There was no rheumatism, and the attack of chorea had followed a fright five days before admission. Here, too, the only important lesion was on the mitral valves—a row of soft, warty vegetations on the auricular face, just within the free margins. (The case is given under Chorea insaniens.)

*Case IV.*—Miss A., aged 27, was admitted to the Johns Hopkins



Hospital December 24th, with violent chorea, of which she died on January 4th. (Case referred to fully on page 23.) The heart was pale and flabby, no signs of old cardiac lesion other than that the bases of the columnæ carneæ of the left ventricle were somewhat fibroid. The edges of the mitral segments were a little thickened, and presented on the auricular faces a row of bead-like vegetations.

*Case V.*—Female, age above 70, admitted to Johns Hopkins Hospital December 2nd, 1893, with chorea of three or four weeks' duration, which was stated to have followed mental worry and disappointment. There was a loud, blowing, systolic murmur. She improved a great deal, but died somewhat suddenly on the 7th of April, after an attack of cardiac dyspnœa. The autopsy showed old mitral valve disease; no fresh vegetations.

As is well known, the mitral segments are the most frequently involved. In forty-three cases they were alone the seat of endocarditis; in thirteen instances the mitral and aortic segments presented vegetations; in three cases the tricuspid segments were involved with the mitral; in two there were vegetations on mitral, tricuspid and aortic valves; and in one case the aortic valves were involved alone. The tricuspid valves may be alone attacked, as in a case reported by Babington.<sup>1</sup> Acute or subacute arthritis had occurred in thirty-one of the cases; in four it was doubtful, and in thirty-seven it was specially stated not to have been present.

Of the fatal cases in which endocarditis was not present in two there was pericarditis, in two chronic mitral valvulitis, in one the heart was fatty.

In the almost constant association with endocarditis fatal cases of chorea stand unique among diseases. The only list of fatal cases of rheumatic fever available with which to make a comparison is that of Guy's Hospital. Of forty-five cases which ended fatally, and in which there had been no previous chronic disease of the valves, the organ was found to be healthy in eight only, in nineteen both endocarditis and pericarditis existed, pericarditis alone in ten, endocarditis alone in eight.<sup>2</sup> The percentage of unaffected hearts in this series is considerably higher than in the chorea cases just given. Endo-pericarditis is very much more frequent in rheumatism, and the proportion of cases of pericarditis is very much greater.

The endocarditis, almost invariably of the simple variety, is shown by the presence of a few small bead-like vegetations just within the

<sup>1</sup> *Guy's Hospital Reports*, Vol. VI, p. 436.

<sup>2</sup> *Fagge's Practice of Medicine*, Third Edition, by Pye-Smith, London, 1891.



margins of the auricular surface of the mitral cusps. They present the usual characters of such structures, and in the cases which I have seen have differed in no respect from the endocarditis met with in rheumatism and in the secondary infections in various febrile disorders. I see no grounds whatever for Dickinson's suggestion that the bead-like vegetations in chorea are not identical with those in other affections. Sansom<sup>1</sup> states that there are two forms of endocarditis in chorea; the ordinary rheumatic valve-thickening, and the beading of the cusps with papillary elevations. Thickening of the edges of the cusps, however, I regard as too uncertain a criterion, and no cases are included in the series which had not actual vegetations. In Case III of my autopsies, a patient of Dr. Molson's, of Montreal, the symptoms and lesions were those of malignant endocarditis. There were numerous large vegetations springing from the auricular edge of the segments in their entire extent. Those attached to the anterior curtain were the largest, and projected considerably beyond the margin of the valve. They were soft, greyish-white in colour, and irregular on the surface. Both spleen and kidneys presented recent infarcts. It was in this case that there was a small embolic lesion in the right lenticular nucleus. In the case reported by Friis, No. 41, and in one of Goodall's cases, No. 32, the endocarditis was ulcerative.

### III.—SUBSEQUENT CONDITION OF THE HEART IN CHOREIC PATIENTS.

It seemed important to determine the subsequent heart history of a considerable number of cases of chorea, since it was clear that in this way alone could satisfactory evidence be obtained as to the influence exerted on it by the primary disease. Stephen Mackenzie has already shown in an examination of thirty-three patients at periods varying from one to five years subsequent to the attack "that indisputable heart disease persisted in 60·6 per cent. of the series of chorea cases examined." Forty-four children examined by Donkin<sup>2</sup> at periods varying from two to twelve years after the chorea eighteen had signs of heart disease. Accordingly in the spring of 1887, with the assistance of Dr. Charles Burr, and in the spring of 1889, with the assistance of Dr. Caspar Sharples, I made an attempt to reach all the cases of chorea which had been in attendance at the Philadelphia Infirmary for Diseases of the Nervous System since

<sup>1</sup> *Lancet*, January 12, 1889.

<sup>2</sup> *Diseases of Children*. London, 1893, p. 302.



1876. In all instances the examination was made *two or more years* subsequent to the attack of chorea. In each case reference was made to the original notes, questions were asked about subsequent attacks and rheumatism, the heart was examined in the recumbent and erect postures, at rest and after exertion, and the notes were dictated at the time of the examination. The results of the examination of the first series, comprising 110 cases, have already been published,<sup>1</sup> and sufficient details given of the cases to indicate the nature of the heart lesion.

Of the 140 cases 98 were in females, 42 in males. The length of time which had elapsed since the attack varied in the cases examined from sixteen years to two years. In the first series more than half (63) of the cases were examined at a period of five or more years subsequent to the attack. The results of the examination were as follows:—In 51 cases the heart was normal; in 17 there was disturbance which might reasonably be regarded as functional; in 72 cases there were signs of organic heart lesion. This gives a remarkable, and I must say at the time, an unexpected high percentage ( $51\frac{3}{7}$  per cent.) of cases in which following an attack of chorea there was definite damage apparent in the heart.

**Normal Cases.**—Of the 51 persons in whom the heart was found to be normal, 15 had three or more, 8 had had two attacks, and 27 a single attack. There was a history of rheumatism in 9 of the cases. In 7 of these the attack was of the acute articular type.

**Functional Cases.**—Of the cases presenting abnormal signs 17 may reasonably be cited as examples of functional disturbance. They were cases without enlargement of the heart and with localized or variable murmurs. Eleven presented soft apex systolic bruits, not propagated to the axilla, and in four varying with the position of the patient. In most of these cases there was also accentuation of the second sound in the second left interspace, a feature upon which, however, no special stress can be laid in young persons, since in them it is by no means uncommon in perfectly normal hearts. Comparison between the intensity of the sounds in the second right and second left interspaces was a point upon which particular attention was paid in examination of the cases. Thus in ten of the normal cases the pulmonary second was distinctly louder than the aortic, and in several instances reduplicated. No stress was laid upon the cardio-respiratory murmurs, which are common in thin-chested young children.

<sup>1</sup> *American Journal of the Medical Sciences*, 1887, I.



In two of these cases with functional disturbance the sounds in the pulmonary area were clear in the erect posture, but when the patient was lying down a systolic murmur was present ; in both the second sound was accentuated, and in one the area of pulsation somewhat increased. In a third case there was a soft, systolic murmur in the second and third spaces in the recumbent position, one with accentuation of the second sound and an apex beat a little outside the nipple line. There may have been in some of these instances organic changes in the valves, but it was deemed best to exclude all those in which the signs were doubtful.

**Cases with Signs of Organic Disease.**—Of the seventy-two cases thirty had had three or more attacks of chorea. The question of rheumatism was carefully investigated in each instance. In twenty-five of the cases, 34·13 per cent., there was a history of acute arthritis, which in seven of the cases had followed the chorea in from one to five years. Comparing the frequency of the articular affection in this group, 34·13 per cent., with that in the total series ; namely 24·2 per cent., or with the group of fifty-one normal cases, in which it was 17·11 per cent., we see illustrated the greater liability to serious heart mischief in the cases with joint complications. We have, however, the much larger proportion, 66 per cent., of cases with positive organic disease examined at a date two or more years subsequent to the attack of chorea, and questioned carefully as to the occurrence of rheumatism, in whom there was no history of the existence of this complication.

*Nature and Seat of the Lesion.*—In a large proportion of all the cases the signs were those of mitral valve disease, usually insufficiency, indicated by the systolic murmur of maximum intensity at the apex, propagated to the left, with evidences of enlargement of the heart, sometimes also by a thrill, and marked accentuation of the second sound in the pulmonary region. The details of a large series of these cases are presented in the paper above referred to. While in a majority of the cases there given the lesion was well compensated, there had been in not a few instances attacks of shortness of breath and palpitation. Three of the cases died with the symptoms of chronic valve disease. In twenty-four instances a mitral pre-systolic murmur was present. In one of these instances (Case LXXXII in the already published series) it was possibly the apex diastolic murmur heard in connection with aortic insufficiency, the so-called Flint murmur. The young man, aged 18, had his first attack of chorea in 1884, a second attack in 1886, and in 1887 a slight



attack of rheumatism. There was a soft murmur at the base in his first attack. In 1887, when examined, there was a loud diastolic murmur of maximum intensity on the sternum, and heard also at the aortic cartilage. At the apex there was a rumbling pre-systolic murmur, localized, and not accompanied with a thrill. One of the cases with mitral presystolic murmur subsequently died. The lad, aged 13 (Case XXXIX of the series published) had his first attack of chorea in 1881, when seven years old. He had a second attack in 1882 and a third in 1884. He never had any joint troubles. Of this, both his father and mother, who were exceedingly intelligent people, were positive. A half-brother has, however, had rheumatic pains in the joints. He had scarlet fever when three years old, but not severely. With this exception he had had no other diseases except chorea. He had, when examined, great shortness of breath and more or less lividity with a distinct pre-systolic thrill in the fourth interspace, a blubbery pre-systolic murmur, also of maximum intensity, in the fourth space, and a loud, blowing systolic bruit transmitted to the axilla. There were signs of considerable hypertrophy of the heart. The autopsy showed extreme stenosis of the mitral orifice with great dilatation of the left auricle and enormous hypertrophy of the right ventricle, with beginning contraction of the tricuspid orifice.

The importance of chorea as a factor in the etiology of chronic mitral valve disease is shown in a striking manner by the examination of these cases. It is unfortunate that it is not always inquired for in the anamnesis of heart cases in the medical wards. It is interesting to note the comparative rarity of the involvement of the aortic valves. There were only four instances of combined aortic and mitral valve disease. This is in accord with the anatomical distribution of the endocarditis, which has been shown to be so much more frequent on the mitral segments.

It may be then stated :

(1) That endocarditis is a very common complication of chorea minor.

(2) That in a majority of such cases the endocarditis is independent of, and is not associated with, acute arthritis, unless indeed we regard the valvular lesion as itself a manifestation of the rheumatism, holding with Bouillaud that "*chez les jeunes sujets le cœur se comporte comme une articulation.*"

(3) That in a considerable proportion of cases, much larger indeed than has hitherto been supposed, the complicating endocarditis lays the foundation of organic heart disease.



## IV.—PERICARDITIS IN CHOREA.

Inflammation of the pericardium in connection with chorea was first described by Bright, who states also that this had been long observed by Guy's Hospital physicians. His statement of the matter is worth quoting: "With regard to the connexion between chorea and inflammation of the pericardium, when called upon the year before last to deliver the Lumleian Lectures at the College of Physicians, I took occasion to state, that for some years I had been persuaded of the existence of such a combination, and little attention has hitherto, as far as I know, been paid to the subject, although the combination of this spasmodic disease with rheumatism has been long recognized. In the very excellent "Syllabus, or Outlines of Lectures on the Practice of Medicine," published at Guy's Hospital, I find, in the edition of 1802, rheumatism distinctly stated as one of the existing causes of chorea; and in later editions, as in that of 1820, I find it stated, that "chorea sometimes alternates with acute rheumatism," but through what organ or by what intervention this occurs is not conjectured.<sup>1</sup> He gives in this paper five cases of pericarditis with chorea and rheumatism; in the first case there was also endocarditis. The Lumleian Lectures for 1836, to which he refers, are not in the *Lancet* of that year, nor can I find that they were published. Bright thought the connection between the two was through the phrenic nerve, which communicated the irritation from the inflamed pericardium to the spinal cord. By far the best account in the literature is that by Sibson,<sup>2</sup> in his exhaustive article on pericarditis. He states that 21 of the 180 cases of acute rheumatism with affections of the nervous system had chorea. Fifteen of those patients with chorea had pericarditis, six had no pericarditis; while fourteen of them had endocarditis; three had no endocarditis, and in three of them endocarditis was probable or doubtful. In 19 of the 73 recent autopsies in chorea which I have collected, pericarditis occurred as a complication, and in seventeen it was associated with endocarditis. In eight of the cases there was a history of acute rheumatism (arthritis).

One case had sub-acute rheumatism, one rheumatic pains, while nine had not had acute arthritis.

Of the nineteen cases eight were under ten years of age, eleven were in the second decade.

<sup>1</sup> *Med. Chir. Soc. Trans.*, Vol. XXII. p. 10.

<sup>2</sup> *Reynolds' System of Medicine.*



In three the pericarditis was old ; in three death is stated to have been caused directly by the pericarditis ; pleurisy occurred in five cases, pneumonia in four ; acute phlebitis in one, and in one acute nephritis.

The following is a very typical case from the records of the Infirmary. I saw it repeatedly with my colleague, Dr. Sinkler,<sup>1</sup> who has reported it.

George W., aged 6, applied at the Infirmary October 29th, 1888, with chorea. Family history was good. He had been well up to February 1888, when, after exposure to cold, he had an attack of inflammatory rheumatism. In March the choreic movements began, and had persisted ; when he came under observation they involved the face, trunk, arms and legs, and the speech was somewhat disturbed.

On November 28th he was admitted to the wards, and under rest and Fowler's solution the movements became less marked.

On January 6th, 1889, he had a severe pain in the umbilical region, and on the 7th in the right side. This persisted throughout the 8th and 9th. On the 9th I made the following note :—"Respiration 18 ; pulse 96, volume small, increased tension. The præcordial region is rather prominent ; the heart's impulse is felt in the third and fourth interspaces, the maximum impulse in line of nipple ; no thrill is felt. The vessels of the neck are prominent, full and pulsating ; præcordial dulness begins above at the third rib ; externally, a little outside of nipple line ; to the right it extends two fingers'-breadth beyond the sternum. On auscultation, a loud systolic murmur is heard at the apex and is transmitted to the scapula. Over the whole præcordial space a loud to-and-fro murmur is heard. The sound is intensified at the base in the third and fourth interspaces ; it is well heard at the nipple and at the ensiform cartilage." Following this the patient had a well-marked effusion, which gradually disappeared, but he was a long time convalescing, and the chorea did not disappear entirely until June. During this time he had no arthritic manifestations, and no subcutaneous fibroid nodules. The temperature throughout the attack was high and sometimes reaching 104°.

<sup>1</sup> *University Medical Magazine*, Vol. II, p. 483.



## CHAPTER V.

## MORBID ANATOMY, PATHOLOGY, AND TREATMENT.

Statistics of Fatal Cases — Changes in the Nervous System — Bacteriology — Pathology — Treatment.

## MORBID ANATOMY.

THERE are no characteristic lesions in fatal cases of chorea. Externally there are frequently bruises and excoriations. In cases of long duration the body is much emaciated, while in acute cases with high fever the appearances are those of a person dead of an infectious disease—the skin is congested, the blood is dark, and the muscles are of a very red colour.

The statistics of fatal cases collected by Sturges and by Raymond have been referred to in the section on the heart in chorea. Of the 73 cases which I have collected since the publication of their figures the general analysis is as follows:—23 were in males, 50 in females; 15 occurred in children of 10 years and under; between the ages of 11 and 15 inclusive there were 17; between 16 and 20 inclusive there were 31; between 21 and 30, there were 6; three cases occurred in the aged, and in one the age was not given. More than double the number occurred in the fourth hemi-decade than in the first two hemi-decades. The ratio of non-fatal cases in these periods is 1 to 4·2.

There was a history of acute arthritis in 29, in three there had been sub-acute rheumatism, and in four the history was defective. Four cases were in pregnant women, and one occurred post-partum. As already stated in the section on the heart, recent endocarditis was present in 62 cases, 85 per cent.; and pericarditis in 19; in two pericarditis alone; in two chronic mitral endocarditis; and in one the heart was fatty. There were in all 66 cases with heart lesions, 90·4 per cent.

*Associated Lesions.*—Pneumonia occurred in nine cases usually lobar in type, but in several cases lobular; acute pleurisy was noted in seven cases; peritonitis in one case; parotitis in two; phlebitis



in two; purulent bronchitis in No. 41; pyæmia (or septicæmia) occurred in Nos. 4, 47, and 57.

Among the incidental lesions may be mentioned hæmatoma of the dura mater (Nos. 16 and 51); acute nephritis (Nos. 31 and 60); gangrene of the foot (No. 32). One patient (No. 25) is said to have died of acute dilatation of the stomach.

It is not easy to say accurately just how many died of the disease itself and how many of the complications.

**Nervous System.**—Naturally special attention has been paid to the condition of the brain and spinal cord. Dana<sup>1</sup> has carefully analyzed the recorded autopsies, of which in only 39 was the condition of the nervous system at all satisfactorily described. Of 19 cases in which careful microscopical examinations were made, in 16 there was intense cerebral hyperæmia, peri-arterial exudations, erosions, softened spots, minute hæmorrhages, and occasional emboli. The changes were most marked in the deeper parts of the motor tract, particularly the lenticular nuclei and the thalami. These vascular changes, perhaps the most constant central lesions of chorea, are essentially the same as those described by W. H. Dickinson in 1876. In Autopsy IV of my series they are very fully described by Berkeley.<sup>2</sup> The peri-vascular spaces were wide, filled with round cells, occasionally imbedded in hyaline masses. The largest transudations were about veins. The adventitia was often covered by small round cells and hæmatoidin *débris*. The muscularis was not hypertrophied. The intima was thickened in spots and presented small swollen nuclei with numerous refractile granules in their substance.

In No. II of the fatal cases of my series, in which the mitral endocarditis was very extensive, there was a spot of embolic softening the size of a cherry in the right lenticular nucleus. It was upon the presence of lesions of this kind in connection with endocarditis that the embolic theory of the disease was suggested and supported by Kirkes, and after him by Broadbent, Tuckwell, and others.

There are two cases reported in which death took place from apoplexy, one by Bevan Lewis,<sup>3</sup> in which there was cerebellar hæmorrhage; in the other by Baxter,<sup>4</sup> a girl of eight years, the hæmorrhage was extraventricular.

Of special histological features the following may be mentioned:—

<sup>1</sup> *Brain*, Vol. XIII, 1890.

<sup>2</sup> *The Johns Hopkins Hospital Reports*, Vol. II, p. 325.

<sup>3</sup> *Medical Times and Gazette*, 1876, II.

<sup>4</sup> *Brain*. April, 1879.



(a) Chorea corpuscles (so-called). These bodies were described by Elischer<sup>1</sup> as sharply contoured, irregular, strongly refractile, concentrically laminated bodies, attached to the blood-vessels in the corpora striata and internal capsule. Jakowenko<sup>2</sup> described them in six cases from Flechzig's clinic. Flechzig himself seemed to lay considerable stress upon their significance.

Wollenberg<sup>3</sup> examined carefully six brains from cases of chorea minor with particular reference to the presence of these bodies, and in addition forty-six brains of non-choreic cases. He concludes that, in some cases of chorea, these highly refractile bodies occur in the neighbourhood of the lenticular nucleus, arranged along the vessels, but that they are in no way characteristic, as they also exist in similar situations in individuals who have never suffered with chorea. He thinks that these bodies represent some calcified organic substance of unknown nature.

(b) Lesions of the pyramidal cells. F. Charlewood Turner<sup>4</sup> has described in the brains of five cases swelling and turbidity of certain of the larger pyramidal cells in the deeper layer of the cortex in the Rolandic region. "The protoplasm is cloudy and dense-looking, and more fully stained than in normal cells, and defined at the bases of the processes, which would indicate a great swelling up of the body of the cell."

Berkeley, in the case above mentioned, could find no special changes in the nerve cells. The peri-cellular spaces were large, but the chief changes found were meningo-vascular, and endo-arterial.

Dana,<sup>5</sup> in a recent case, which, however, was associated with chronic lepto-meningitis, found hyaline degeneration of the pyramidal cells.

(c) Lesions of the medulla and spinal cord. No characteristic changes have been met with in these parts. Berkeley found many foci of minute hæmorrhage in the pons, especially among the fibres of the pyramidal tract. The blood-vessels showed the same changes as in the cortex. "The nuclei on the floor of the ventricle, and more anteriorly the olivary bodies, the internal fibres of the vagus and hypoglossus, as well as the transverse fibres, were examined with great care and patience, but without finding the slightest pathological

<sup>1</sup> Virchow's *Archiv*, Bd. LXIII.

<sup>2</sup> Reference in *Neurologisches Centralblatt*, Bd. VIII.

<sup>3</sup> *Archiv für Psychiatrie und Nervenkrankheiten*, Bd. XXIII, 1891, p. 167.

<sup>4</sup> *Pathological Society's Transactions*, Vol. XLIII, 1892.

<sup>5</sup> *American Journal of the Medical Sciences*, 1894, I.



change in them." In the spinal cord the multipolar cells were mainly perfect in protoplasm, nucleus and prolongation. In canine chorea, an affection entirely distinct, however, from chorea minor in man, H. C. Wood has described changes in the ganglion cells. "When the animal was killed in the very beginning of the attack, the cells showed no change; a little later the only alterations in the cells were the very frequent absence of the nuclei, the failure of granulations in the protoplasm, the loss of power to take staining fluids, and rarely the occurrence of sharply-defined vacuoles. Then the processes began to drop off; and finally it was found that the places of the cells were occupied by irregular, globose, crumpled-looking masses, without sharp outline, and taking carmine staining very faintly. No granulations, no nuclei, no processes, were apparent. These masses represent the cells in the last stages of degeneration."<sup>1</sup>

Berkeley,<sup>2</sup> in a dog which had had chorea for between five and six months, found the cells of the spinal cord in all respects perfectly normal.

Triboulet,<sup>3</sup> however, who has made a very careful examination of the spinal cord in cases of canine chorea, states that he has confirmed the observations of Wood, and finds very marked lesions in the cells of the anterior horns. He agrees, however, that the canine chorea is an entirely different disease from that seen in man.

**Bacteriology.**—Attempts have been made to isolate micro-organisms in chorea. From what we know of the invasion of the body by bacteria, we should expect to obtain cultures in the acute febrile cases. Such a series of observations as that of my colleague Prof. Welch<sup>4</sup> on the organisms found in cultures made from 180 autopsies shows how cautious one should be in judging of the etiological value of the presence of certain forms.

The observations which have been made so far on chorea are not at all satisfactory. Naunyn found a cladothrix on the meninges and in the endocardial vegetations.

In the fatal case from my wards reported by Berkeley, the cultures made from the blood in the left ventricle, from the vegetations on the valve, and from the parotid gland, showed an abundant growth of the staphylococcus pyogenes aureus.

<sup>1</sup> *Nervous Diseases and their Diagnosis*, Philadelphia, 1887, p. 155; also *Therapeutic Gazette*, 1885.

<sup>2</sup> *Johns Hopkins Hospital Reports*, Vol. II, p. 337.

<sup>3</sup> *Paris Thesis*, 1893.

<sup>4</sup> *Middleton-Goldsmith Lecture*, 1894.



Triboulet<sup>1</sup> deals extensively with the possible infective character of chorea, and has made a number of cultures, in two fatal cases finding the staphylococci; in another instance he found staphylococci in the blood during the febrile attack.

The most exhaustive research on the subject is that by Pianese<sup>2</sup> from the Pathological Institute of the University of Naples. He claims to have isolated a bacillus from the nervous system of a choreic patient, which he was able to cultivate successfully. Animals inoculated died with muscular twitching and convulsions, and from these animals the same bacillus could be obtained in pure cultures from the central nervous system. His work, a copy of which he very kindly sent me, is illustrated by beautiful plates showing the cultures and the micro-organisms in the tissues.

Dana<sup>3</sup> has recently reported a case, not, I should suppose, of Sydenham's chorea, in which in the meninges a diplococcus was found resembling the diplococcus lanceolatus. A few other observations are found in the literature, as Richter,<sup>4</sup> who found cocci in the blood, and Donkin,<sup>5</sup> who found rod-like bodies in the tissues. Altogether the only observations which lay claim to anything like completeness are those of Pianese, and they await confirmation.

#### PATHOLOGY.

A satisfactory presentation of the pathology of chorea cannot be given in the present state of our knowledge; and now, at the close of the century, as at the beginning, in the time of Bouteille and Bernt, there are many problems awaiting solution. While our clinical information has widened and has become more accurate, the quotations from these old worthies, which I give on page vi, express the judgment of our own day on the obscure nature of the malady.

It is not my intention to discuss the various views which have been broached; the student and the specialist will find them fully considered in the larger works on diseases of the nervous system, particularly in that of Gowers, and in the recent monograph of Sturges (2nd edition). I have thought it best to take up the recent suggestion that chorea minor is an infectious disorder, and to present some of the points which are urged in favour of this view.

<sup>1</sup> *Paris Thesis*, 1893.

<sup>2</sup> *La Natura Infettiva della Corea del Sydenham. Ricerche anatomiche sperimentali e cliniche del Dottore Giuseppe Pianese.* Naples, 1893.

<sup>3</sup> *American Journal of the Medical Sciences*, January, 1894.

<sup>4</sup> *Western Lancet*, Vol. XII.

<sup>5</sup> *Medical Times and Gazette*, 1884.



The severer types of Sydenham's chorea have all, the milder forms some, of the features of an acute infectious disease, and were these graver types alone seen, no one would question its position ; but in the slighter varieties the suggestion seems strained, while the psychical element in causation, often so striking, is very strongly opposed to any such theory. The rôle of infection in producing diseases of the nervous system is recognized as of growing importance. The infections may be thus grouped :—

(a) Tuberculosis and cerebro-spinal meningitis, in which the infective organisms are present in the lesions.

(b) Diphtheritic paralysis, tetanus, and possibly hydrophobia, in which the toxic materials are alone present.

(c) Syphilis, of which neither the organism nor its toxine is known, and which has the remarkable dual action, the one proximate, inducing gummata and arteritis ; the other remote, causing sclerosis.

(d) Then comes a group of diseases in which infection is possible, but for which satisfactory evidence is still lacking :—some forms of acute neuritis, acute polio-myelitis, acute encephalitis, and here some authors would place chorea minor.

The following points favour the view that chorea is an acute infection :—

1. The influence of age, sex and season, in which it presents the peculiarities common to infectious disorders.

2. The clinical course of the severer forms, in which not a single feature is wanting of a typical infection. The disease presents, as occurs in infections, all gradations between the mild form with scarcely an unpleasant symptom, to cases of such intensity, as in the one reported by Cook and Beale, that death may occur in 130 hours.

3. The post-mortem appearances. Endocarditis, the most constant change, upon the significance of which not sufficient stress has been laid by writers, is one of the most distinctive lesions of an infectious disease. Varying in frequency in different disorders, and produced by a variety of organisms, still, so far as we know, it does not occur apart from the conditions resulting from what we designate as an "infection." In the opinion of many good observers its presence is really enough to stamp the nature of chorea. Other lesions, such as pericarditis, pleurisy, parotitis, septic inflammation, point in the same direction.

Intimately associated with this question is that of the relation of chorea to acute rheumatism. Are its symptoms merely manifesta-



tions of the rheumatic poison, or does the arthritis bear the same relation to chorea as the joint inflammation to gonorrhœa, or to cerebro-spinal fever?

One of the "vulgar errors" of the profession has been the abuse of the term rheumatism, which, almost in the same way as chorea, has been used to cover a multitude of totally different affections. Acute rheumatic fever, an acute infection of unknown origin, is a well-characterized and only too common disease; but all cases of acute poly-arthritis are not rheumatic fever. There are many infectious diseases, in which arthritis develops as a secondary complication—scarlet fever, typhoid fever, cerebro-spinal fever, dysentery, septicæmia and gonorrhœa.

Gonorrhœa is an acute local infection, often accompanied by arthritis, and sometimes complicated with intense pericardial and endocardial inflammation. No one now believes that this arthritis is identical etiologically with acute rheumatism. The pain, the redness, and the swelling may be the same, but the general behaviour, the irregular localization, the greater liability to suppuration, the protracted course, the resistance to the salicine compounds, all point to a different cause.

There is an acute affection of the nervous system directly comparable with chorea in this very matter of acute arthritis. In epidemic cerebro-spinal meningitis, arthritis occurs in a variable percentage in different outbreaks; thus, in the recent one reported upon by Flexner and Barker,<sup>1</sup> "nearly twenty per cent. of the severe cases suffered from complicating joint affections, the knees being most frequently affected, the elbows, wrists, ankles following in frequency in the order named. The effusions were peri-articular as well as articular, and the joints were swollen and red, resembling closely the appearance of those in acute rheumatic arthritis; indeed there were cases which, had it not been for certain initial symptoms indicating the meningeal process, could have easily been diagnosed as nothing more than attacks of rheumatism." They note also that the cerebral symptoms appear to be favourably influenced by the joint affections. There is scarcely an argument used by the strong advocates of the rheumatic nature of chorea which could not be applied most forcibly in favour of the rheumatic origin of epidemic cerebro-spinal meningitis. Some indeed would come with telling force, such as the almost invariable association of the disease with inclement weather and changeable seasons.

<sup>1</sup> *American Journal of the Medical Sciences*, 1894, I.



The nature of the virus of rheumatic fever is unknown. Several forms of micro-organisms have been found associated with acute poly-arthritis, and in one instance, with all the features of an acute rheumatic fever, the micrococcus lanceolatus was present. A careful study of the bacteriology of the disease has recently been made by Professor Sahli,<sup>1</sup> of Bern. He regards the acute onset, the self-limited course, the complication and the frequency of arthritis in other infections, the similarity in many respects of the disease to a septicæmia—all as supporting the view that it belongs to this group. He has found in some cases an organism identical with the staphylococcus citreus, but of very low virulence. In connection with this point Flexner and Barker make an interesting suggestion in discussing the joint lesions of cerebro-spinal meningitis. "Nevertheless it is probable that acute articular rheumatism will, ere long, be proven to have no *etiological unity*, just as has already been proven for the inflammations of the serous membranes generally. It seems probable that the entrance of pyogenic organisms of different varieties into the circulation, under circumstances which are inconsistent with the development of the phenomena of a general septicæmia, may give rise to inflammations in some one or more of the serous membranes of the body—be it meninges, pleuræ, pericardium, peritoneum, or joint surface, the particular ones attacked depending on certain peculiarities either in the virulence of the invading organism or in the lessened resistance at the moment of the serous membrane implicated."

Evidently we are as yet upon the threshold only of our knowledge of the essential cause of either acute rheumatism or chorea. In both disorders there are facts highly suggestive of an infective nature, but more than this cannot be said at present. The relationship of the two diseases remains unsolved. If, as some would have it, chorea is only one of the rheumatic states, we have to stretch beyond recognition our conception of the disease, now, in the absence of a knowledge of its etiology, necessarily characterized by its symptoms. Very probably the cause of chorea will be found to be a poison allied to, but not the same as, that of rheumatism. On this question of the infective nature of the disease Gowers very justly remarks that when the causation of a disease is complex it often happens that at different periods attention is directed too exclusively to one of the elements of its production.

While, on the one hand, the course of the milder forms, and the

<sup>1</sup> *Deutsches Archiv für klinische Medicin*, Bd. LI.



not infrequent association with fright, favour the view that the disease is a simple neurosis ; on the other, the course of the severer forms, the arthritis, and the almost constant presence of endocarditis in the fatal cases, speak for an acute infection.

It is interesting to note that the evidence in favour of the infectious character of canine chorea is now very strong.<sup>1</sup>

The SITE of the primary change in chorea minor has been much discussed. That the condition found in fatal cases gives no satisfactory information on this point is evident, since, basing their arguments on these findings, authors have claimed in turn the cortex, the basal ganglia, and the cord as the seat of the disease. As in the consideration of the nature of the disease, the aggravated cases give us such important information, so with respect to the seat of the primary change, the intense psychical phenomena indicate clearly a cortical lesion. The alterations in character, and the mental features of the malady in less severe forms, also suggest involvement of the higher centres. The cessation of the movements during sleep, and the frequent occurrence of hemi-chorea, are usually urged as favouring this view. Of course, irregular movements of muscles may be produced by irritation in any part of the motor path, but "the motor impulses that excite the muscles pass to the spinal cord from the motor region of the cortex. It is here that movements are arranged, and if they are disarranged and the disorder proceeds from the brain, we naturally refer it to a disordered action of the cells of the cortex" (Gowers). The part played by the spinal cord is probably subsidiary in the chorea of man. H. C. Wood urges that the ganglion cells in the whole cerebro-spinal system suffer, and he has adduced experimental evidence to prove that in the dog at least the cord is chiefly involved. But, as already remarked, canine chorea is a different disease from chorea minor in man. From time to time cases are reported which suggest the spinal cord as the seat of the lesion, but, as in those recorded by S. Weir Mitchell and Burr,<sup>2</sup> they are usually not cases of chorea minor.

#### TREATMENT.

Of the conditions favouring the development of the disease three are important and may be guarded against :—

(a) Brain and eye-strain at school. These are perhaps the most important, and a child who has had one attack of chorea minor

<sup>1</sup> Triboulet, *Paris Thesis*, 1893.

<sup>2</sup> *Journal of Nervous and Mental Diseases*, 1890, XV.



should be prohibited from competing for prizes in class work, and should have the lessons carefully regulated.

(*b*) In children of families subject to rheumatism very special care should be taken, particularly in the spring months, that there is no unnecessary exposure to cold and damp.

(*c*) The nutrition of the child should be maintained at a maximum, and on the first appearance of anæmia, iron and arsenic should be used. These are measures which can be urged upon a mother whose child has already had an attack.

The most important elements in the treatment of the attack itself are rest and seclusion. The child should be put to bed and kept there until the movements have ceased. This may seem an unnecessarily severe procedure for a disorder so simple, apparently, as chorea minor, but the very mildest cases are not without danger, and it is probable that the liability to heart complication is considerably diminished by complete rest. As the movements diminish the child may be moved to a lounge. The remarkable influence of this procedure in allaying the severity of movements is often seen in hospital practice, and a case which in the out-patient department has seemed of extreme severity, has, at the end of two or three days, been, without any medication, changed to one of comparative lightness. In private practice it is a measure usually resented by mothers, and may be very difficult to carry out. I have often insisted, where the family could afford it, upon the presence of a special nurse.

Next to rest, perhaps the most valuable measure is seclusion. Usually the children affected are very bright and intelligent, and correspondingly sensitive, and the consciousness of their affliction is a constant source of worry and irritation. The child should not see many persons, and when possible should be in charge of either the mother or an intelligent nurse, whose main occupation should be to read to the child and keep it amused.

In the severer cases with great jactitation it becomes a question of protecting the child from injury. The mattress should be very soft, and all neighbouring hard objects should be guarded with soft cushions and pillows. It is sometimes well to cover the elbows, knees and ankles with cotton-wool. In the terrible cases of chorea insaniens the liability to bed-sores is very great, and a water-bed may be necessary. It is well to remember that in children the movements may be such that the child is thrown out of bed, an accident which I have known to occur several times in hospitals, in which the beds are as a rule very unsuitable for choreic patients. It is



sometimes better in a severe case to make up the bed on the floor in a corner of the room.

The diet should be abundant and nourishing. Sometimes the question of feeding may become a very difficult one, when the head is in a constant state of movement. An ordinary baby's bottle with a nipple and a piece of rubber tubing may then be necessary. Milk, broths, nourishing soups, and eggs can be given in this way. Except in the very light cases, it is best to have the child fed by a nurse.

Systematic massage and various movements may be employed when possible. In aggravated cases it is out of the question, but when the movements are not very violent thorough inunction with oil or cocoa-butter may be practised daily for half an hour. The full warm bath is often very grateful, and I have seen the movements diminish in the bath and after its use. Of course great care must be taken to prevent the child knocking itself when in the tub. In the severer cases of chorea insaniens the cold bath or the cold pack may be employed, particularly if there is high fever.

There are no remedies which directly control the course of the disease, and which can be called curative. Among remedies which modify the symptoms and appear to do good the following are the most important.

*Arsenic.*—For years this remedy (recommended by Thomas Martin,<sup>1</sup> of Reigate) has been employed in chorea, and it is probably the safest to give as a matter of routine practice. It is well to begin with small doses, three minims, after meals, of the Fowler's solution well diluted. This may be increased two minims every third or fifth day until the child is taking twelve or fifteen minims three times a day. Martin says that he "began with five drops and increased one drop every day until it might begin to disagree with the stomach or bowels," which was usually when a dose of fourteen drops was reached. The dose was then diminished and continued at ten drops for six weeks. The general condition of the children is often much improved by this drug, but it is extremely difficult to say whether it controls in any way the choreic movements. I do not myself think that it has very much influence upon them. When large doses are given the effects must be carefully watched, and the arsenic should be stopped on the first indication of any toxic symptoms, such as vomiting, diarrhoea, itching of the eye-lids, œdema, or skin eruptions. Pigmentation of the skin is occasionally met with; a more serious effect is neuritis, of which there have been

<sup>1</sup> *Medico-Chirurgical Transactions*, Vol. IV, 1813.



recently several cases reported. Considering the rarity of this toxic action upon the nerves, the cases in which it occurs must be regarded as instances of idiosyncrasy. Thus Marshall reports<sup>1</sup> a case of neuritis and pigmentation in a boy of six years of age, who had taken Fowler's solution of arsenic,  $\text{m} \times \text{t. d.}$ , for about five weeks.

The zinc compounds were much used during the first half of this century.

Of other remedies *cimicifuga*, antipyrin in full doses, that is from twenty to sixty grains in the day, chloral, exalgine, sulphonal, and physostigmine have all been warmly recommended. Recently H. C. Wood has urged the use of quinine in large doses, and has reported very favourable results.

Certain features and complications require special treatment. In the cases with arthritis the joints should be wrapped carefully in cotton-wool, and opiates may be needed to allay the pain. The salicylates may, of course, be tried, but in the arthritis associated with chorea we rarely see the prompt and satisfactory results so constant in acute rheumatism. Should pericarditis or endocarditis supervene, the ice-bag may be kept applied to the præcordia. It allays the cardiac excitement, and in the former condition checks, I believe, the tendency to exudation.

In the chorea insaniens hydrotherapy should be thoroughly tried, either in the form of the wet-pack or the bath. In order to apply the pack or put the patient in the bath it may be necessary at first to give chloroform, which in any case may be used with safety when the jactitation is excessive.

Good results have been reported by Bastian and by Gairdner in these cases by large and increasing doses of chloral hydrate, keeping the patient continuously under its influence. The bromide of potassium may be combined with it. In these cases, too, the heart becomes feeble, and there is great prostration, for which alcohol should be given freely. Gee,<sup>2</sup> who has reported six recoveries in seven cases, thus sums up his experience of chorea insaniens: "to prevent bed-sores, to keep the patients clean and to feed them are the most important parts of the treatment."

In the chorea occurring during pregnancy the same general treatment should be carried out, particularly the quiet and seclusion. When the jactitation is extreme the patient may be kept under the influence of chloroform. The induction of premature labour is recommended in very severe cases.

<sup>1</sup> *Lancet*, 1890, I.

<sup>2</sup> *St. Bartholomew's Hospital Reports*, Vol. XXII.



## CHAPTER VI.

## CHOREIFORM AFFECTIONS.

*Habit Spasms, Tic.*

- I. Simple Tic—Habit Spasm or Habit Chorea—Generalized Tic. II. Tic with Imperative Ideas, &c. III. Complex, co-ordinated forms of Tic. IV. Cases of Noisy Spasm of Respiratory Muscles.

I PROPOSE in this section to give a brief description of those forms of spasmodic contraction of the muscles which are known as habit spasms, habit chorea and tic. It is best, perhaps, to employ the latter term, extending the Anglo-American usage so as to embrace not alone the local spasms of the facial muscles in children, but the more extended co-ordinated movements described by the French as *tics convulsifs* and *tics co-ordonnés*. Their consideration here is appropriate on account of the frequency with which the cases are confounded with chorea minor.<sup>1</sup>

Litre's definition of tic, "A local and habitual convulsive movement, a contraction of certain muscles, particularly those of the face," has been much extended by the Salpêtrière school; and under this term is now embraced a series of disorders of musculation, simple or co-ordinated, and with or without psychical manifestations. Guinon, in his elaborate article in the *Dictionnaire Encyclopédique*, gives the following definition:—"An habitual and conscious convulsive movement, resulting in the contraction of one or more of the muscles of the body, reproducing, most frequently in an abrupt manner, some reflex or automatic action of common life." The action may be controlled, or at least modified, to some extent by an effort of the will; and in this, as in the reproduction of a movement, reflex or automatic in character, it differs from the

<sup>1</sup> Charcot very aptly says (*Leçons du Mardi*, 1888-89, p. 464):—"Entre le tic et la Chorée il y a un abîme: ne l'oubliez pas, car il s'agit là affections auxquelles on donne quelquefois, bien à tort, le même nom et dont le pronostic est bien différent." And again:—"Sans doute, nosographiquement, les tic et la Chorée représentent bien, comme je vous l'ai dit, deux affections radicalement distinctes."



involuntary, bizarre, and much slower contraction of the muscles in chorea minor.

As the varieties of tic pass insensibly into each other, it is difficult to make a satisfactory classification of the cases. I shall describe them in the following groups:—*Simple tic*, localized or general; *la maladie des tics convulsifs* of the French; the co-ordinated tics; and lastly I shall speak of some forms of spasm of the respiratory muscles, as allied to the tics.

### I.—SIMPLE TIC.

#### *Habit Spasm. Habit Chorea.*

The spasms may be localized or general.

**Localized Tic.**—This, one of the commonest disorders of movement, begins usually in young persons, and may persist through life. The spasms are confined to a single muscle, a group of muscles, or a group of associated muscles. The muscles of expression are most often involved, and in the common parlance of the profession *tic* means facial spasm. The idiopathic facial spasm of adults differs in many respects from habit spasm of the facial muscles. It is rarely seen until after the 40th year, is more common in women, the muscular contraction has not that quick, electric-like quality, but there are more often tonic and clonic spasms. The various forms of habit spasm are too well known to require description; suffice it to say that the contraction may involve a single muscle, as the orbicularis palpebrarum on one side, or a group of the muscles of expression. Many forms are of trivial importance, but the more severe type, in which nearly all the muscles of the face are affected, and in which, during speaking, the depressors of the chin and the tongue muscles are thrown into action, constitutes one of the most distressing of the minor ailments of life. In many of these cases the affection seems to begin as a childish trick, particularly the blinking of the eyes, and the quick, rapidly repeated act of sniffing. Though not usually grouped with tic, the various forms of spasmodic wry-neck really belong to the same category of muscular disorders. The tic may be confined to the platysma muscle, as in a case which I saw a few months ago with Dr. H. M. Thomas. Habit spasm of the shoulder muscles is not uncommon, and is often a movement associated with a grimace or the act of winking.

Simple tic is not so often seen in the muscles of the arms and



legs, either in single muscles or in groups ; but in the former there may be quick movements of flexion and extension or of rotation, or of flexion and extension of the fingers.

In the legs simple tic is less common. The most striking is the "springhalt tic," in which in walking at irregular intervals the leg is flexed rapidly on the thigh, sometimes with an associated movement of the muscles of one arm or of the face.

The following is a remarkable case of long-standing tic of the muscles of the right leg :—

*Tic of the Muscles of the Right Thigh lasting for Thirteen Years, of late occasional Spasmodic Contractions of the Right Hand ; no Explosive Utterances.*

D. C., aged 44, lawyer, seen April 25th, 1893, complaining of a nervous twitching of the muscles, particularly those of the front of the right leg. He has always been a vigorous, healthy man ; comes of very good stock, and there are no nervous diseases in the family. He has been a moderately heavy smoker ; has taken alcohol daily ; a little more lately than usual, on account of worry about his condition.

Thirteen or fourteen years ago, at a time when he was working very hard, he first noticed, in bed, just before going to sleep, that the muscles of the right thigh would twitch, and sometimes contract strongly enough to flex the thigh on the abdomen. At other times there would be a little tremulous creepy feeling beneath the skin of the thigh. It worried him very much and would at times keep him awake. There was no pain, but he was a good deal upset by it, and even dreaded the idea of going to bed. For a long time it was confined to the muscles of the thigh, but during the past few years, particularly if he had been very hard at work and much worried, a sensation would pass up the right side, and the arm and hand would jerk. Lately, if very nervous, or if hurried when writing, or if he was watched, the muscles of the hand would jerk a little, and once or twice the hand shook so that he had to stop writing. Otherwise he has been quite well ; there have not been any sensory disturbances, except an occasional uneasy feeling about the right leg or a little numbness or tingling in the right arm. Once or twice he has had vertigo. As a younger man he suffered a good deal from migraine, but has not had it at all lately. There have been no mental symptoms ; and no explosive utterances. The only acknowledgment of any special nervous sensation was sometimes feeling a little ill at ease and out of sorts when in company.



The examination was entirely negative ; there was no wasting of the muscles of the right leg ; sensation was perfect, and there were no changes in the reflexes. There was no indication of involvement of the nerves of the leg or of the spinal cord.

The physical examination of the thoracic organs was negative. The patient was urged to quiet his mind on the subject, as he had had apprehensions that it would be a progressive trouble and cause paralysis, and told to rely rather on bathing and the cold pack before going to bed, than on the bromide and on alcohol. The twitching at night has evidently been serious enough to cause him a great deal of mental distress, and has been associated with a dread of going to bed and going to sleep, for which reason he has taken more bromide and alcohol than was good for him.

**Generalized Tic.**—There is a very interesting group of cases in which the tic manifested in sudden, electric-like jerkings of the muscles of the trunk and extremities, causing a start which shakes the patient for an instant, and passing leaves him motionless and tranquil. The cases occur both in children and in adults. In the former the condition has been described as electric chorea<sup>1</sup> from the shock-like character of the movements. It is, however, not at all uncommon in adults, particularly in women, and the condition may persist for years. Very many of the cases have been described under the head of chronic chorea minor. The following are abstracts of histories of illustrative cases in adults :—

*For Four Years Electric-like Spasms of the Muscles of the Trunk and Limbs. Twitchings of the Right Corrugator Supercillii and Right External Rectus.—Delusions.*

Susan B., aged 32, admitted November 24th, 1891, complaining of jerkings and general nervousness. The family history is good ; her father died of heart disease ; the mother and sisters have no nervous disorders.

When eight years old she had typhoid fever, during which there was a transient speech disturbance. She grew to womanhood without any special troubles, and, with the exception of minor ailments, remained well until about four years ago, when she had

<sup>1</sup> The name Electric Chorea has also been given to Dubini's disease, probably an acute infection, met with chiefly in Lombardy, characterized by an onset with pains in the back and neck, and then contractions of the muscles, like those following electric shocks. They begin in the fingers, and spread over the body. Paresis of muscle groups occurs, and finally coma and death.



acute Bright's disease with dropsy, from which she did not recover for a year and a half. During the convalescence the present trouble began with jerkings of the body and limbs, which have persisted ever since. She has never had any convulsions. Lately she has become suspicious of her friends, and has had delusions. The attacks, which come on at any time, consist in electric-like contraction of the muscles of the trunk, very abrupt and quick, passing off in a moment, and not moving her from the chair, or the place in which she may be standing. In bed, however, they will lift the back momentarily. Sometimes they succeed each other rapidly for a minute or two, but she may pass an hour or part of a day without them. There are no facial grimaces and no movements of the hands or feet. After she had been under observation for a few weeks she began to have twitching of the right corrugator supercillii, and the right external rectus contracted at intervals, causing an outward jerking of the eye-ball. On account of the increasing mental trouble the patient was transferred to an asylum.

*Electric-like Jerkings of the Trunk Muscles and Extremities for many Years ; Chronic Tuberculosis.*

A. B., aged about 27, was under treatment in Ward C for pulmonary tuberculosis of long standing. She had had for many years (she did not know how long) spasms in the muscles, sometimes in those of the face alone, most frequently involving also those of the body and extremities. She had been treated by many physicians without avail, and had gradually learned to accept the condition as hopeless. Of late, since the onset of the lung trouble, the movements have been less marked. She was a small, delicate-looking woman, with chloasma, and bulbous fingers and toes. While talking the face muscles would occasionally twitch, particularly those of the eyes, but the spasm was not at all excessive. At intervals of a few minutes or longer she gave an electric-like start, in which also the legs and arms seemed to participate, but she would not drop an article she was holding at the time. These jerkings did not disturb her very much, and she had become accustomed to them. They ceased during sleep. She was not hysterical.

*Sharp, sudden Spasms of Muscles of Trunk and Extremities of a Year's Duration, Epilepsy, Cessation of the Movements during Typhoid Fever.*

S. F., aged 21, admitted to ward G with involuntary jerkings of



the muscles and epilepsy. Her mother is of unsound mind. She has been well and strong, but for several years had had epileptic attacks. A year ago she began to have twitchings of the muscles of the body and limbs, and for this she sought relief.

She was a well-nourished girl, a little dull-looking and pale. Every few moments there were quick, lightning-like contractions of the muscles of the arms and trunk, strong enough to lift the arms, but she would not drop objects from the hand. They were increased by excitement, and seemed worse when she was in bed. The legs also participated, but not to the same extent as the arms. In the intervals between the jerkings there were no twitchings. The face was not affected. The movements ceased during sleep. About sixteen days after admission she developed typhoid fever, during which the movements ceased entirely, to recur gradually during convalescence.<sup>1</sup>

*Sprain of Ankle, Remarkable Choreiform Movements of the Trunk Muscles on the following Day. Neuritis of Nerves of Right Leg.*

Lieut. X., aged 31, in the service 10 years; always healthy and strong; dyspepsia occasionally.

Family history good; father died at the age of 71; mother died in childbirth. Brothers and sisters well and strong; no nervous troubles in family; no history of any spasms or nervous troubles.

He had not chorea as a child; but since about the age of fifteen he has always been a little tremulous when excited. He has never noticed anything but this tremulousness. In 1886 he had muscular rheumatism after exposure; pain in the shoulder and the legs below the knees. In October, 1890, he sprained the right ankle in jumping from a horse which was falling backwards. The ankle was bandaged, and he walked the next day with a stick; but the joint was sore and swollen. Five or six weeks after he sprained it again, and the pain was so severe that he had to have a hypodermic injection. He has been lame ever since. The day after the ankle was sprained he had remarkable nervous twitchings, which seemed to be a jerking in the muscles of the trunk and abdomen. He seemed to feel as if he was under tension, and then, not being able to resist it, the jerking would come on. They were not painful and not frequent, occurring only four or five times a day. For

<sup>1</sup> The case is fully described in the *Report on Typhoid Fever, Johns Hopkins Hospital Reports*, Vol. IV.



three or four months they continued to trouble him, but at the Hot Springs he was better, and had no attacks. In New York, during the summer of 1891, while under treatment for wasting of the right leg, there was no return of the spasms. He returned to duty in October 1891. He was not able to do any riding, and gradually the nervous jerkings returned. They were not painful but were very unpleasant, as any one near by could notice them. He never had any jerking of the face muscles or of the hands. The spasms were confined entirely to the trunk muscles. There was no pain in the back at this time. He remained at the post until January 1892, when he was ordered to detached service with the Militia. He was very much better for a few months, though the ankle was still weak and painful. He was on duty until the 1st of November, and had no "jerking spells" except at the latter part of August. Altogether he was very much better, and in June was able to ride occasionally, and gave up walking with a stick.

In November he was not so well, and the ankle troubled him a good deal. He was in Washington from November 1st until March 1893, under treatment most of the time for the back, which had become painful, and the leg. He had no jerking spells until February. In March he went to North Dakota, and there felt well until the 10th of April, when he had pain in the ankle, and the nervous jerkings recurred, and the riding caused pain in the back. The pains extended down the right leg, but were centred chiefly in the lumbar region. At one time they became so severe that he could not dress himself or turn. He was on the couch all day for a week, then was up and down for a couple of weeks, with occasional, but not severe, jerkings.

From the 16th of May he has been in the house with a good deal of pain in the back, and when sitting up has had to have a pillow behind him.

*Present Condition.*—He is a well-built, well-nourished man of six feet one inch in height, and looks very robust. When stripped he looks a man of fine physique, with well-developed muscles, but the right leg is decidedly smaller than the left. This is very evident on inspection before and behind. There does not appear to be any special difference between the thighs. The ankle is not swollen; there is no wasting of the muscles of the feet; pressure is a little painful at the posterior part of the inner malleolus, and for a couple of inches in this region in the course of the posterior tibial nerve. The patient can perform all the movements, and there is no weak-



ness of any special group of muscles. The lumbar muscles are equal in volume, and the spine is straight. All the twisting and bending movements of the back are performed without any pain. He complains of a little pain and tired feeling in the lumbar region, and there is a little sensitiveness on deep pressure in the course of the right sciatic nerve. There are no sensory disturbances in the area of distribution of the sciatic nerve.

The knee-jerks are normal ; not exaggerated on either side.

Examination of the other organs is negative. The pupils are equal, and respond to light.

The patient does not give one in any way the idea of a neurasthenic subject.

He remained under observation for about two months, and with massage and electricity the local conditions improved rapidly. The jerkings, as he calls them, occurred now and then. I only saw them once, when a sort of exaggerated shiver seemed to pass through him, and moved the trunk and shoulders, but not the head or extremities. He says they were very transient and went through him like a flash.

## II.—TIC WITH COPROLALIA, ECHOLALIA, &C.

(*Maladie de la Tic Convulsif. Gilles de la Tourette's Disease.*)

In the second group may be placed the cases which present, in addition to the motor disturbance, certain remarkable features, as the explosive utterance of certain words or sounds, and a mental state characterized by the existence of fixed ideas. Particular attention has been paid to this form by Charcot and his pupils, and it has been made the subject of a special memoir by one of them, Gilles de la Tourette,<sup>1</sup> whose name is now often given to the affection in France.

The following cases will illustrate fully the chief features of the affection.

### CASE I.—*Tic for Eight Years ; Coprolalia ; Echolalia.*

Mary —, aged 13 years, applied at the out-patient department Johns Hopkins Hospital, July 10th, 1890, and was under observation there until September 16th, when she was admitted to ward G.

<sup>1</sup> *Archives de Neurologie*, 1885. In the *Leçons du Mardi* of Professor Charcot, 1887-88, pp. 65, 105, 294 ; and 1888-89, pp. 13, 464, will be found a full discussion on many interesting cases.



Her mother brought her to the hospital on account of irregular involuntary movements and curious barking sounds.

The family history is good. Her mother is a bright, intelligent woman, a German by birth, who has had ten children, none of whom have been affected as is this girl—the third child. There is no tendency to mental disease in the family. The birth of the child was normal and there is no history of convulsions in infancy. She has had scarlet fever, but has not had rheumatism.

Since her fifth year she has been subject to involuntary jerking movements of the arms and head, which vary very much in intensity, sometimes better, sometimes worse, and they have usually been called by the doctors chorea. They have not interfered with her development or her education. She has not yet menstruated. For the past year she has been making curious sounds; beginning by saying “hah” very frequently. Sometimes she would bark like a dog. She would also call out the names of people, and if she heard a new name she would be apt to repeat it.

Her condition on admission was as follows:—A bright, intelligent child; well educated, writes nicely, takes an interest in her books and has evidently been ambitious at school. The right arm occasionally twitches and the head jerks. There are no grimaces, but on several occasions she seemed to mimic movements of the face. Every now and then she calls out “hah,” “Bridget,” or “stools”; or says in sharp, clear tones, “bow, wow.” There are no disturbances of sensation, and the special senses are unimpaired. Examination of the heart and lungs is negative; the thyroid gland is slightly enlarged.

Throughout the latter part of July and August attempts were made to treat the case by hypnotic suggestion, at first with success, but subsequently without any improvement.

On September 8th her mother wrote the following letter, which illustrated a new phase of the child's malady:—

“Mary makes use of words lately that make me ashamed to bring her to you, or to take her out of the house; it is dreadful; such words as —, —, —, etc. She was always a modest child, and it almost kills me for to hear her use such words.”

Her mother was asked to bring her again, and was told that this was really a part of the affection, and, like the movements, involuntary in character. The child seemed more depressed, had lost flesh and, her mother said, had changed mentally. She was very obstinate, and almost invariably did what she was told not to do, and had



threatened to take poison. She will say the bad words aloud or mutter them to herself.

On admission to the hospital she was placed in a room by herself, kept in bed, and encouraged in every way to cease making the sounds and to stop the use of the bad words. During the first two weeks she improved very much. The movements were reduced in frequency, and sometimes during my visit they would not be noticed at all. They most commonly affected the right arm, which, with the hand, was drawn up in a sudden electric-like jerk. The head and neck would jerk simultaneously or alone. Sometimes there was combined movement of the neck and chest-muscles. The involuntary expressions of which she made use were those mentioned above; a sharp bark was the most frequent sound, which, from its ringing quality, could be heard at a considerable distance.

She improved and was allowed to get up, and another patient was placed in the room with her. This seemed to excite and worry her, and shortly afterwards the barking sounds became much more frequent, occurring every one or two minutes, and she complained of great soreness of the muscles of the chest and abdomen. The movements, however, did not increase. She was again placed in seclusion and in bed, and again improvement followed, but she still barked and did not give up entirely the use of bad words.

She is a docile, intelligent child, and seems anxious to get well. She has kept a diary, which displays no special peculiarity. She writes verses, which are not worse than those usually composed by girls of her age.

CASE II.—*Tic of the Muscles of the Face and Neck ; Fixed Ideas ; Arithmomania.*

A. B., aged 13, seen Sept. 6th, 1890. She is an only daughter in a family with marked neuropathic taint. The father died insane; the mother is a high-strung, nervous woman.

The child is well grown, and well nourished, though rather stout for her age. She is very bright and intelligent, and perhaps has not had as much control as was good for her. For a year or more she has had occasional twitchings of the muscles of the face and neck, noticeable in the quick sudden elevation of the eyebrows, or in movements of the platysma muscles. They have not been severe, and for days it may not have been at all noticeable. There have been no spasmodic movements of the arms or legs. The condition has not interfered in any way with her growth or development. She



is very fond of outdoor exercise, particularly of riding on horseback. A short time after the onset of the twitchings it was noticed that she began gradually to have all sorts of queer notions and practices, many of which persisted for some weeks or months, and were then changed for others not less anomalous. Some of her vagaries are as follows, nearly all being modifications of the fixed idea known as arithmomania. Before getting into bed at night she lifts each foot and taps nine times on the edge of the bed. After brushing her teeth she has to count one hundred. For a year at least she has always entered the house by the back door, protesting that she never can enter by the front door again. Lest her mother should prevent her getting in by the back door, she for months carried the key herself. On reaching the door she knocks three times on the edge of the window near by, and three times on the door before unlocking it. She will not under any circumstances button her shoes. For a long time she would not pronounce the name of anyone, but would spell it, and if she wished for anything at the table she would spell the word, but not pronounce it. In drinking water she will take a mouthful, then put the tumbler down, turn it once or twice and repeat this act every time she drinks. She would not brush her hair except at the extreme tips, and it is only under the strictest compulsion that she will allow the hair on the top of the head to be combed. Before putting on clean under-clothes she has to count so many numbers that there is a great difficulty in getting her to make the change, except under the strongest threats from her mother.

The patient was sent to the country under the care of her aunt, who was urged to control and train the child.

A special interest attaches itself to this case, inasmuch as the patient has recovered completely. I have seen her on several occasions within the past three years, and she has grown into a fine young woman; both the tic and the mental symptoms have disappeared.

CASE III.—*Convulsive Tic, with Echolalia, Coprolalia, and Délire du Toucher.*

Delia L., aged 12, sent by Dr. Goldsborough of Cambridge, Md., complaining of involuntary movements and of loss of control over speech.

*Family History.*—She comes of good stock without neurasthenic taint. Father and mother are living and well. The mother seems



a sensible woman and is not at all nervous. There are two sisters and two brothers living and healthy.

*Personal History.*—She has been very healthy; she has had measles and whooping-cough; but neither rheumatism nor muscular pains. She is of an excitable temperament, and very bright mentally and ambitious at school. She gets very excited at play, and lately has been using a tricycle.

About a year ago the mother noticed that she winked her eyes quickly at times, and had jerking movements of the head. As they were slight at first not much attention was paid to them. Subsequently she had sudden, quick twisting movements of the body. The arms did not jerk, but she had occasional jerking movements of the legs. Since the summer the movements of the head and trunk have increased a good deal, and come on at intervals of a few minutes or so, and are much aggravated by excitement. She continued very well physically.

In August, 1893, a new feature developed; she would at intervals shout out words of all sorts without being able to prevent it, particularly such words as "murder," "fire," and at times obscene words which she picked up from hearing boys use them in the street. These would sometimes be called out very loudly with an associated movement of the head. She also repeated words or short sentences which she heard, and after reading a street sign, she would often call it out in a loud voice several times, or if she met a person and heard his name she would repeat it loudly. About the same time she began to touch objects, and would stoop down and touch the floor, or touch certain things in the room, particularly the lamps. She also would frequently stoop down and knock with her knuckles on the floor a certain number of times, or would rap on the table.

Her general condition has kept good. She has often been excited and wayward, but her appetite is good and she sleeps well.

*Present Condition.*—She is a well-grown girl for her age. The hair and eyes are dark, and the expression very bright. She sits quietly for a time and talks without any embarrassment and quite readily. Occasionally she will wink rapidly with her eyes, and elevate the eyebrows. Every few minutes the head jerks with a sudden, quick, lateral, and forward movement, evidently with a contraction of the muscles of the thorax as well, as the trunk also moves slightly. These occur at irregular intervals, and are rather more frequent under excitement. Every few minutes there is a



sudden explosive utterance. During the examination there was no special word, but an indefinite sound, often quite loud. Frequently during the examination she would touch objects, apparently without self-consciousness ; thus she would stoop and touch the floor, and she touched on several occasions the tip of the pen with which Dr. F. A. Smith was taking notes. There were no movements of the hands or of the legs.

The physical examination was negative. There were no sensory disturbances, and the reflexes were normal.

CASE IV.—*Tic with Subjective Auditory Hallucinations.*

Eliza D., coloured, aged 53, seen January 13th, 1894. She is a married woman ; has been healthy and well as a rule, and so far as can be gathered has had no serious illnesses, or any mental disturbances until the onset of her present trouble. The menopause occurred five years ago. At this time she was much troubled with hot and cold flushes and began to have a twitching of the left shoulder, and it is for this, which has continued, that she seeks relief.

Patient is a well-nourished woman, a little excited and garrulous. Every few minutes the left shoulder is lifted and the head drawn to the left. The movement is rather slow and does not affect the arm and hand. When she is standing up it evidently involves the trunk muscles, as the body is rotated and makes a half turn as the shoulder is drawn down. This may occur without any rotation of the head, but sometimes they occur together. The twitching has continued almost uninterruptedly, and, she thinks, has of late become aggravated. But what troubles her most seriously are certain sounds which she hears, as if people were talking to her. She never herself makes any explosive utterances, but during the movements she hears distinct sounds or noises, which she says are only present when the movements occur. It is impossible to get a good account of the nature of these noises, whether they are words or merely sounds. Some time ago she had also hallucinations of sight, but these have not been present lately.

The special features of the affection may now be considered.

(a) **The Involuntary Movements.**—These vary from trivial, slight spasm of the facial muscles, as in Case II., simple tic, to the most extraordinary bizarre movements, involving all the muscles of the body. The muscles of the face are most commonly affected, and there is a sudden, rapid closure of the eyelids, elevation of the



eyebrows, or a quick, lightning-like movement of the muscles of expression, producing an unnatural-looking smile. As in simple habit spasm, a somewhat frequent form is the quick, rapid sniffing of air. Instances have been described, too, in which there were rapid movements, alternately opening and closing the mouth. The chin may be suddenly depressed, and a frequent movement is the rapid contraction of the platysma, moving the skin of the neck and lower part of the face and over the shoulder. The movements may be limited to the face, one side or the other, but more commonly other muscles are affected, and a sudden spasm of the sternomastoid may draw the head quickly to one side, and at the same time there is a facial grimace. A not uncommon movement is a sudden jerking upward of the head, with a rapid lateral motion. Affection of the muscles of the shoulder girdle is common on one or both sides, and with or without movements of the arm. The movements of the upper extremities are very varied. There may be, as in Case IV., with rotation of the trunk, a drawing backwards of the arm; or, as in Case III., slight jerking movements, sometimes of pronation and supination, or of flexion and extension. Other movements may be made of a purposive nature; thus the movements, as in Case III., of definitely touching an object, which is common, or of scratching any part of the body, or the hand may be frequently placed before the mouth as in coughing, or both hands may be rubbed together as if in great glee.

Isolated movements of the trunk muscles are not so common, but the salaam movements may be present in this affection.

Isolated movements of the lower extremities are rare, but the leg may be abruptly extended, or, what is more common, there is either, as the patient stands or walks, a sudden spring-halt, due to contraction of the flexors of the legs and of the thigh.

And lastly, there are instances on record in which the movements are of a most unusual and astonishing character; thus a patient whom I saw at the Salpêtrière, and whose case is described by Gilles de la Tourette, would run rapidly for a short space, then touch the knee, and perform the most remarkable movements with the arm. As a rule the patients have no difficulty whatever in carrying out the movements of the will, that is, the ordinary voluntary movements. They write, feed themselves, etc., without any trouble, though sometimes in a patient affected with serious tic of the arm there may be some difficulty in writing.

The movements are much influenced by emotional causes; thu



in the presence of strangers, and when under observation, as during the examination, they may be greatly exaggerated. When the patients are quietly at home, or isolated under treatment, the movements may be quickly reduced. In some instances any sudden sound may at once cause a series of rapid irregular contractions. They cease always during sleep, and in several cases it has been noted that an intercurrent fever has checked the movements entirely. In some instances a powerful exercise of the will may restrain to a great extent the movements, but in young children the anxiety in a voluntary attempt to restrain them sometimes causes an aggravation.

There are curious associated actions with the tic; thus a lad under my care at the Infirmary, who had facial tic, was in the habit of rapidly grasping the middle finger between the teeth, biting it quite hard, and at the same time pressing with the index-finger the tip of the nose. So frequently had the action been performed that thick callosities had been produced on the skin of the second phalanx of the middle finger. Such actions are difficult to separate from the simple tricks which children practise, as in the case of Hartley Coleridge, who when a boy was in the habit of biting his arm; and I remember seeing an instance a few years ago of a young girl recovering from chorea minor(?) who when she took anything into her hand had the curious trick of first smelling and then blowing upon it.

(b) **Involuntary Cries.—Coprolalia.—Echolalia.**—Cases I. and III. illustrate very well the character of the explosive utterances which form so characteristic a phenomenon in this affection. These may be nothing more than the exclamation of “ah!” “ahem!” or “oh!” associated in each instance with an involuntary movement. As in Case I. the same may have a somewhat barking quality, two sounds succeeding each other quite rapidly. In Case III. there was the involuntary utterance of all sorts of words, such as “murder!” “fire!” or any name which she had heard, and which seemed to take her fancy, would be repeated throughout the day. Not only would the girl repeat words, but short sentences, and after reading a sign on the street she might repeat it aloud many times.

A remarkable feature, in some instances, is the irresistible tendency to repeat words or names which have been heard, and which Gilles de la Tourette has designated by the term echolalia. The word or phrase which the patient has heard may be frequently repeated; thus on the day on which Case I. was brought to the hospital, she



heard the word nurse frequently, and repeated it for some time. The patient may often echo words which she has said herself. A distressing feature, and which gives to the affection one of its most annoying characters, is the habit which the patients have of using bad words, designated by the word *coprolalia*. Though these are, as a rule, involuntary, yet the child may be to a certain extent restrained by the presence of strangers; thus in Case I. the girl was in the habit at home of using shocking words, to the great distress of her mother, but during examination, and, indeed, during her stay in the hospital, she very rarely employed them. In Case III. this feature of the malady gave the greatest possible distress to the relatives. Thus with a shrug of the shoulder, or with a facial grimace a young child may horrify her parents or those about her by saying "God damn," or "Jesus Christ," or using words of the most obscene character! They are often used on the street, and may sometimes get the poor victim into difficulties, as in a young lad, whose case is reported by Gilles de la Tourette, who called out with his tic, *couillon*. He was thought by some other boys to be addressing them, and the poor coprolalic was given a good thrashing.

Not only may the patients mimic or echo words, but certain movements may also be mimicked at once, to which habit Charcot has given the name *echokinesia*. Any movement which is made is at once imitated, whether it be a facial grimace or a movement of the hands or of the body. This feature is marked in the affection known as Latah, seen among the Malays, and which, with the jumpers of Maine and the Myriachit of Russia, appears to be a variety of tic.

(c) Many cases present remarkable *mental features*, which, with the explosive utterances and coprolalia, complete the picture. They come under the group of the imperative ideas,<sup>1</sup> *obsessions mentales* of the French. These fixed ideas, as they are also called, may be a very troublesome manifestation, taking very varied forms, among the most common of which is the agoraphobia, or the dread of walking in a large open space; the *folie pourquoi*, or insanity of doubt, in which a patient incessantly demands the reason for the performance of even the simplest acts of every-day life; the *onomatomania*, a curious mental state, in which a word or name constantly recurs, and the impulse to repeat it is irresistible. Case II. illustrates well the obsession known as arithmomania. Before attempting to perform

<sup>1</sup> For full discussion of these, see Article by Dr. D. Hack Tuke, *Brain*, summer number, 1894.



the common, every-day actions of life, this child had the irresistible impulse to count a certain number of times, or had to tap a certain number of times with her hand or foot. Another common manifestation is the imperative impulse to touch certain objects. This was particularly well seen in Case III., and in her it seemed to be almost a spontaneous unconscious action. This *délire du toucher* in children is often a mere trick, and may persist to later life, as in the well-known case of Dr. Johnson. In adults, associated with forms of tic, it constitutes one of the most interesting forms of street pathology; thus, I have seen a man who, as he walked along, would go beneath the windows of a house, swing his hand slowly two or three times, and then touch a portion of the window. It is a very common trick in boys, and usually of no special moment. George Borrow makes use of it in an interesting manner in his "Lavengro," describing his imperative impulse to touch the top of a certain tree. In his case it was associated with an idea that the performance of the action would ward off some evil to his mother.

The *prognosis* of this affection is very uncertain. Usually the cases last for many years, and Charcot and his pupils regard the outlook as very unfavourable, Guinon stating that it is, as a rule, incurable. In Case I. the condition had lasted for eight years, and she did not improve at all under seclusion and treatment in the hospital. Case II., on the other hand, made a complete recovery, and both the arithmomania and the involuntary movements have disappeared. The coprolalia may persist an indefinite time, as in the case of the Marquise de Dampierre, who involuntarily used shocking language on most inappropriate occasions from his earliest youth to the age of ninety.

### III.—COMPLEX CO-ORDINATED TICS.

Under this heading may be grouped a number of forms of habit movements, differing from ordinary tic in the more complex character of the action performed, which may be one of every-day life, but one which is repeated without obvious cause and which can be controlled or arrested by an effort of the will.

(a) Many tricks and habits are of the nature of this co-ordinated tic, as the action of a distinguished President of the Royal College of Physicians, who in writing stopped at every few words and looked intently at his finger tips. The tricks of children, head-nodding (though not always), thumb-sucking, and rocking in bed (a habit which may persist to adult life), are allied actions. Some of these



forms I have already mentioned in the last section, particularly the case of the child recovering from chorea minor (?), who on taking anything in the hand first smelt and then blew upon it, and the boy with facial tic, who bit his finger hard and pressed his nose at the same time with the index-finger.

Possibly the interesting affection of children known as head-nodding, and which has been described so fully by Gee,<sup>1</sup> Hadden,<sup>2</sup> and Peterson,<sup>3</sup> belongs in this group. In a case at the Infirmary for Diseases of the Nervous System, a child, aged two-and-a-half years, would sit on the floor by the hour, playing with a few toys, and nodding the head every few moments; not a simple up-and-down motion, but with a rotation from left to right and right to left. The child had also lateral nystagmus and was feeble-minded. Some of the cases have followed injury, usually in early life, but not at birth. The nystagmus and the feeble-mindedness point, in some cases at least, to organic changes, yet both Gee and Hadden state that the prognosis for recovery is fair. The condition is to be differentiated from *epilepsia nutans* in children.

Another strange disorder of allied nature is the "head-banging" in children, of which cases are described by Gee in the same volume of the Bartholomew Hospital Reports. In one case, a child of five, the habit had lasted two-and-a-half years. Asleep or awake, while in bed, the child would turn over and bang the head violently into the pillow, repeating the act five or six times. In another case the act was repeated for two or three hours at a time. No other symptoms were present. In one of the cases a younger brother also caught the trick.

(b) An extended series of these co-ordinated movements are met with in feeble-minded children. Balancing, rotation of the head from side to side, and the striking of the chin violently against the chest are extremely common habits. The repeated beating of the forehead or the face with the hand or fist is another trick, frequently seen in imbeciles, to which Rubinowitch has given the name of Krouomania. Many of these movements are rhythmic, particularly the balancing, the nodding, and the rotation. Noir,<sup>4</sup> from Bourneville's division of the Bicêtre, has described many cases of tic of various kinds in the blind, particularly the rapid movements of the fingers before the eyes. In other instances the movement may be one of jumping, in which the patient makes a series of leaps.

<sup>1</sup> *St. Bartholomew's Hospital Reports*, Vol. XXII.

<sup>2</sup> *Lancet*, 1890, I.

<sup>3</sup> *Medical News*, 1892, I.

<sup>4</sup> *Étude sur les Tics*. Paris, 1893.



In others, again, he performs in orderly sequence a series of actions, such as stooping from the chair, lying prone on the floor, raising the hands above the head, &c., all of which are repeated from time to time.

At my visits to the Institution for Feeble-minded Children at Elwyn, while studying the hemiplegic and epileptic cases, one room in the Hillside division had always a special attraction. In it was a remarkable collection of the unhappy victims of "irresistible musculation." One had not crossed the threshold before the hand was grasped by a bright-eyed, well-knit little fellow, sharp and active as a fox-terrier and restless as a wolf, an imbecile, with the motor centres abnormally active, and with a facial tic, displayed in a lightning-like contraction of the muscles of expression. Hemiplegics with mobile spasm, poor William B., a case of double athetosis, and epileptics, apparently engaged our attention, but we watched in reality a curious Astec-like idiot, known as the "Dervish." In a few minutes he would rise from the floor, balance for a moment, take two or three gentle, sweeping rotations, then poise himself and begin a series of the most extraordinary gyrations, moving but slightly from one spot. At first the rotatory movement was slow, and readily followed, but soon the speed increased, and with arms out and clothes flying, and form and features almost unrecognizable, he span round like a humming-top until he dropped exhausted.

(c) In this group of co-ordinated tics may be placed those cases of extraordinary and bizarre movements repeated at intervals for perhaps a long period of years, sometimes, but not always, associated with imperative ideas or explosive utterances, as described in the preceding section. The remarkable case reported by Weir Mitchell<sup>1</sup> of a man who for years had a sort of pendulum spasm, in which, unless at perfect rest in the recumbent posture, the left arm would strike the side in a regular order, at the rate of from 150 to 160 times in a minute, is an exaggerated example of this form of tic.

#### IV.—CASES OF SPASM OF THE RESPIRATORY MUSCLES.

There is a series of cases in which a recurring spasm affects the muscles of respiration and phonation, and the muscular contraction is accompanied with more or less noise, either a snuffle or hiccough during *inspiration*, or some noisy utterance or explosive sound during *expiration*. These constitute the two varieties. While

<sup>1</sup> *American Journal of the Medical Sciences*, 1876, II.



many of these cases are hysterical, in others no features save the respiratory spasm are met with.<sup>1</sup> The following are illustrative cases :

CASE I.—*Loud Inspiratory Noises following a Fit of Anger.*

Rebecca T., a Russian girl, 20 years of age, came to the dispensary on November 5th, complaining of what she called hiccough.

As regards her family history no neuropathic tendencies could be traced, and she herself has shown no special signs of nervousness. Excepting an attack of typhoid fever two years ago she has always been a healthy and strong girl. Her menstruation has been regular to October of this year. A year ago, following some family trouble, she left her home and came to this country.

Her present disease began shortly after a fit of anger about the middle of October. She then began to produce peculiar spasmodic sounds, which at times she utters in quick succession, while at others she remains perfectly quiet for as long as two or three hours. She has also had crying spells. For some years she has had severe headaches, which, since the onset of the present illness, have become more intense and more constant. In the waiting-room of the dispensary she was quiet, and had been so, we were told, on the street. Immediately upon entering the examining room, however, she began to produce these gasping sounds.

The following note was made in the ward :—The patient is a well-nourished girl, not anæmic, and there is no fever. The examination of the thoracic organs is negative, and nothing abnormal is detected in the abdominal viscera. There are no tender points along the spine. Patellar reflexes are quite active ; they may even be called exaggerated. The most striking peculiarity about the patient consists in the production of loud, gasping noises, occurring during inspiration and often prolonging it. They come on at irregular intervals, and are usually quite loud, but vary in intensity and frequency, increasing in both respects during observation. Expiration is normal, and during it no sounds are ever produced. During sleep they are absent and respiration is quiet. Sometimes the sounds bear a resemblance to a hiccough. During the crying spells she rarely shed tears, but merely produced sounds in rapid succession which resembled sobbing. If the door is suddenly opened the noises at once begin, while they may be absent for some length of time

<sup>1</sup> The best discussion on the different forms is by Charcot in the *Archives de Neurologie*, 1892.



during quiet conversation, even if she has been extremely noisy immediately before. Sometimes the sounds are short and produced in quick succession, or six or more long-drawn, very loud inspiratory sounds may follow each other. During her stay at the hospital a rise of temperature was noticed, especially on the first, third and fourth days, reaching  $100^{\circ}$  twice, once  $99.5^{\circ}$ .

In this case hysteria was very probably the underlying condition, though there were no disturbances of sensation or of sight. In a very similar case in the wards last winter the hysterical characters were pronounced.

It is difficult sometimes to determine the exact origin of these explosive sounds; thus, in the following case the patient insists that the noise which she produces is preceded by a rumbling roll very low down in the abdomen, and both she and her sister speak of it as noisy belching, but from the character of the noise which she describes it is more likely to be produced in the respiratory passages.

#### CASE II.—*Noisy Explosive Sounds.*

Miss F., aged 24, seen with Dr. Bosley, May 23rd, 1893, complaining of a very remarkable noise which she makes at intervals. The patient has always been well and strong. She has had measles, scarlet fever, and whooping-cough. Eight years ago after an alarm of fire in a theatre she lost her voice for three months. She has, however, enjoyed very good health and has not been specially nervous. She has not had hysterical spells; *i.e.*, of laughing and crying, and has not been very emotional. She lost her father a year ago, and since that time she has been living a very quiet life. Her mother is an exceedingly quiet, reserved woman; not at all nervous. The other members of her family are perhaps a little excitable and nervous.

About the last of March, one Wednesday evening, hearing a knock at the door, and expecting some girl friends, she ran to open it. She had a "caramel" in her mouth at the time. Instead of finding the persons she expected there was a young man of her acquaintance, and not liking to be found with her mouth full, she swallowed the bon-bon suddenly. She felt no bad effects at the time, but that night had cramps in the abdomen. These continued the next day, and she had diarrhoea and passed a little blood. The following night too she had quite severe cramps. Evidently, from what her sister said, she was a good deal alarmed and uneasy about herself. On Saturday morning she began to make a remarkable



noise, something like a belch, but very much louder, and this has persisted ever since. She describes it as a rumbling roll which comes from low down in the abdomen, passes up the stomach and comes out at her mouth as an explosive loud noise. She can tell beforehand by uneasy feelings in the abdomen, but finds it impossible to stop its onset. She can put her handkerchief quickly to the mouth and deaden the noise. It occurs from sixteen to eighteen times in the day. It is more frequent in the morning after she gets up. It is much less when she is at rest, but turning in bed is apt to produce it. No fluid comes up with it; sometimes a little acid. Occasionally there are two or three smaller sounds following the first loud explosion. The sister describes the noise as "terribly" loud, and it can be heard several rooms away; frequently on the street it is enough to make people start. Diet does not appear to have much influence. It has been a good deal worse lately on peptonized milk. Curiously enough it is not worse when she is in company, or not rendered worse by emotion. Thus her doctor has never heard it, and in the half-hour which she spent at the consultation with me the sound was not once made.

The patient is a well-built, medium-sized young girl, looks bright, and the colour good. She has, however, lost five or six pounds, and her sister says she does not look at all well. The physical examination is entirely negative; there are no other hysterical features in the case; the stomach is not dilated. During the examination she stated that frequently the abdomen would swell and become too tight for her waist-band. The right kidney is not palpable.

She was so insistent that the sound came from the abdomen that I advised her to have the stomach washed out once or twice a week, but it had no influence, psychical or otherwise.

CASE III.—*Tic of the Deglutition Muscles, with a slight Clicking Noise; subsequently Facial Tic.*

Laura H., aged 11, a very bright-faced, nervous-looking child, applied at the Dispensary April 5th, 1894, complaining of a curious movement in her throat. An uncle had Parkinson's disease, and the mother had chorea. The child has had typhoid fever, chicken-pox, and measles.

When three years old she had an operation for strabismus performed. She is a very bright girl, and is advanced at school.

Two weeks ago her present trouble began quite suddenly with a peculiar motion of her throat, which continues constantly during the



day, and disappears only when she is asleep. She has no other twitching, and complains of nothing else. At first there seemed to be a peculiar noise in the throat, which has, however, now almost ceased. There is no history of any explosive utterance.

On the 6th I made the following note :—A bright-eyed, clear-cut, intelligent-looking child ; no movement of facial muscles ; slight strabismus of left eye. At first nothing was noticeable, but in a few moments rhythmical movements began in the neck, which were practically those made in deglutition, and associated with elevation and depression of the hyoid bone, and of the thyroid cartilage. These parts are drawn up and down rhythmically, causing a wavy movement beneath the skin, such as occurs in the act of swallowing. They are repeated sixty-eight times in a minute. Drinking water makes no apparent difference. During the movement there can be felt a sort of click, and when she holds her head back a slight noise is heard, also like a click, as though the cartilages were rubbing over one another. The heart-sounds are somewhat rapid, and the first is reduplicated at the apex.

The movements in the throat gradually ceased. I saw her June 26th, and she said she had not had them for two weeks, but she has now a well-marked facial tic, involving the muscles of the eye-brows and the right orbicularis palpebrarum.

Various forms of this respiratory spasm are in reality quite common. Terribly exaggerated instances, among the most distressing of all afflictions to the friends and relatives, are encountered. On the 30th of June, 1884, I saw brought into Professor Wagner's clinic at Leipzig a most remarkable case of this kind ; a young girl, aged fifteen, who had always been healthy, but who had worked very hard at school. Some time about the end of March she awoke one night with difficulty in breathing, and ever since has had the remarkable respiratory spasms and cries to be described. They have persisted the entire day, ceasing only with sleep, and as the child has taken but little food, she has wasted to a skeleton. Before the patient was wheeled into the clinic, the noises which she made could be heard at a great distance. They consisted of a loud and very intense *inspiratory* cry, sometimes preceded by three or four short, jerky inspirations. Then following the cry there was a deep, hoarse, very loud expiratory sound. These followed each other with remarkable sequence, the child sitting up in bed and swaying to and fro. The examination of the lungs was negative ;



there was, of course, extreme dryness of the throat. I abstract the following from the notes which I made on the occasion: "Professor Wagner put his thumbs in the child's mouth and depressed the lower jaw forcibly, and urged her to breathe quietly and gently. The movements ceased almost immediately, she took a deep inspiration, lay back in the bed, and breathed quite quietly, told her name, and said that she had been much overworked at school. She took a glass of water and seemed quite quiet and natural." I did not hear the subsequent history of the case.

A few years ago Dr. Gapen of Omaha brought a phonographic cylinder on which was recorded a cry of a somewhat similar nature, uttered by a young girl for many months. It was loud enough to be heard at a distance of several city blocks.

No doubt the *ptussis canina* of old writers, in which the patients uttered short explosive sounds like the bark of a dog, though in many cases an expression of hysteria, belongs in this category; possibly, too, some of the instances of the barking cough of puberty.



## CHAPTER VII.

## CHRONIC PROGRESSIVE CHOREA.

*Huntington's Chorea.*

Historical Note—Report of two Families—Special Symptoms—Morbid Anatomy  
—Diagnosis.

**Historical Note.**—The affection was first described by physicians of the State of New York, in parts of which it appears to have existed for very many years. In the first edition of Dunglison's "Practice of Medicine," published in 1842, on page 312 of the second volume, he gives a letter from Dr. C. O. Waters, of Franklin, New York, in which an affection is described as prevailing in the south-eastern sections of the State, where it was known as the "megrims"—"consisting of a spasmodic action of all or nearly all the voluntary muscles of the system—of involuntary and more or less irregular motions of the extremities, face, and trunk." The throat muscles sometimes participated, as evidenced by the clucking sound. The disease was stated to begin in adult life, was markedly hereditary, and was incurable. I do not see, as has been stated, that Waters had noticed the association with dementia.

The next description, also from the State of New York, is found in the *American Medical Times* for December 19th, 1863, in an article on Chronic Hereditary Chorea, by Dr. Irving W. Lyon, House Physician, Bellevue Hospital. No mention is made of the section of the State in which the cases were found. He states that the disease was almost exclusively confined to certain families, which were popularly designated the "megrin families." The hereditary transmission was fully recognized in the community, "and the people among whom it occurs believe this to constitute its only legitimate method of propagation, and acting accordingly, have repeatedly been known to interdict marriage alliances between their children and those believed to be tainted with the megrim diathesis under the severe penalty of social ostracism." Dr. Lyon



gives instances in which cases occurred through five generations, but gives no details, and there was no recognition of the mental deterioration.

In 1872, Dr. George Huntington, of Pomeroy, Ohio (now of La Grangeville, N.Y.), published in the *Medical and Surgical Reporter of Philadelphia*, April 13th, an every-day sort of paper on chorea minor, at the conclusion of which he described a form which he called hereditary chorea, met with at the eastern end of Long Island, New York, and well known to both his father and to his grandfather, who had practised in that locality. As I have never seen Huntington's description reprinted in full, I give it here, calling attention merely to the really graphic account he has given in a few paragraphs of the salient features of the disease.

"There are three marked peculiarities in this disease:—1. Its hereditary nature. 2. A tendency to insanity and suicide. 3. Its manifesting itself as a grave disease only in adult life."

1. "Of its hereditary nature. When either or both the parents have shown manifestations of the disease, and more especially when these manifestations have been of a *serious* nature, one or more of the offspring almost invariably suffer from the disease, if they live to adult age. But if by any chance these children go through life *without* it, the thread is broken, and the grandchildren and great-grandchildren of the original shakers may rest assured that they are free from the disease. This you will perceive differs from the general laws of so-called hereditary diseases, as for instance in phthisis, or syphilis, when *one* generation may enjoy entire immunity from their dread ravages, and yet in another you find them cropping out in all their hideousness. Unstable and whimsical as the disease may be in *other* respects, in *this* it is firm; it never skips a generation to manifest itself in another; once having yielded its claims, it never regains them. In all the families, or nearly all in which the choreic taint exists, the nervous temperament greatly preponderates, and in my grandfather's and father's experience, which conjointly cover a period of seventy-eight years, nervous excitement in a marked degree almost invariably attends upon every disease these people may suffer from, although they may not when in health be over-nervous."

2. "The tendency to insanity, and sometimes that form of insanity which leads to suicide, is marked. I know of several instances of suicide of people suffering from this form of chorea, or who belong to families in which the disease existed. As the disease progresses



the mind becomes more or less impaired, in many amounting to insanity, while in others mind and body both gradually fail until death relieves them of their sufferings. At present I know of two married men, whose wives are living, and who are constantly making love to some young lady, not seeming to be aware that there is any impropriety in it. They are suffering from chorea to such an extent that they can hardly walk, and would be thought, by a stranger, to be intoxicated. They are men of about fifty years of age, but never let an opportunity to flirt with a girl go past unimproved. The effect is ridiculous in the extreme."

3. "Its third peculiarity is its coming on as a grave disease, only in adult life. I do not know of a single case that has shown any marked signs of chorea before the age of thirty or forty years, while those who pass the fortieth year *without* symptoms of the disease, are seldom attacked. It begins as an ordinary chorea might begin, by the irregular and spasmodic action of certain muscles, as of the face, arms, etc. These movements gradually increase, when muscles hitherto unaffected take on the spasmodic action, until every muscle in the body becomes affected (excepting the involuntary ones), and the poor patient presents a spectacle which is anything but pleasing to witness. I have never known a recovery, or even an amelioration of symptoms in this form of chorea; when once it begins it clings to the bitter end. No treatment seems to be of any avail, and indeed now-a-days its end is so well known to the sufferer and his friends, that medical advice is seldom sought. It seems at least to be one of the incurable."

Subsequent to Huntington's description but very little is found in the literature until 1884, when Ewald<sup>1</sup> called attention anew to the question, and reported two cases. In this country Clarence King<sup>2</sup> (of Machias, N.Y.) reported another family. Then followed in rapid succession in a few years many observations by Peretti,<sup>3</sup> Huber,<sup>4</sup> Hoffmann,<sup>5</sup> Lannois,<sup>6</sup> and others. In 1889 appeared the monograph by Huet,<sup>7</sup> in which the historical details and the histories in full of the reported cases to that date are given. Since this publication there have been numerous additional articles, of which the following are the most important:—

<sup>1</sup> *Zeitschrift f. Klin. Med.*, Bd. 7, supplemental Heft, 1884.

<sup>2</sup> *New York Medical Journal*, 1885, Vol. XLI.

<sup>3</sup> *Berliner Klin. Wochenschr.*, 1885, p. 824.

<sup>4</sup> *Virchow's Archiv*, Bd. 108.

<sup>5</sup> *Virchow's Archiv*, Bd. 111.

<sup>6</sup> *Revue de Medicine*, 1888.

<sup>7</sup> *De la Chorée Chronique*. Paris, 1889.



Suckling,<sup>1</sup> Diller,<sup>2</sup> Bower,<sup>3</sup> Biernacke,<sup>4</sup> Hay,<sup>5</sup> Jolly,<sup>6</sup> Remak,<sup>7</sup> Cirincione and Mirto,<sup>8</sup> Mendel,<sup>9</sup> Sinkler,<sup>10</sup> Reynolds,<sup>11</sup> Greppin,<sup>12</sup> Dreves,<sup>13</sup> Schlesinger,<sup>14</sup> Phelps,<sup>15</sup> Lannois and Chapnis,<sup>16</sup> Ruffini,<sup>17</sup> Kronthal and Kalischer,<sup>18</sup> Oppenheim and Hoppe,<sup>19</sup> and Menzies.<sup>20</sup>

**Symptomatology.**—I will in the first place give the history of the cases which have been under my personal observation in two families,<sup>21</sup> which illustrate well the hereditary nature of the malady.

#### FAMILY X.—*First Generation.*

A. B., an Englishman, married C. D., a native of — County, State of —, and had issue eleven children. A. B. died aged 87, and his wife aged 85. Neither of them, so far as is known, displayed any mental or bodily peculiarities. Two of the eleven children died choreic and demented.

Of the other children, two of the girls married N.'s. One died aged 75, leaving children, all of whom are in good health; the other, Mrs. N., still lives, aged 77, and has healthy children; George died aged 70, a bachelor; Sarah died aged 50, of typhoid fever, without issue; William died aged 76, leaving a large family, none of whom have shown any symptoms of the disease; Mary died of an acute illness, aged 55, leaving healthy issue; Jane died aged 70, leaving a family, none of whom are affected; two other daughters died maidens, well advanced in life. The two affected children were James and Margaret.

James, the first to become affected, began to exhibit remarkable muscular irregularities before he was 40. Dr. Ellis writes: "I very well remember, in my earliest youth, his grotesque movements, exciting unusual attention, and I fear more ridicule than sympathy.

<sup>1</sup> *British Medical Journal*, 1889, II.

<sup>2</sup> *American Journal of the Medical Sciences*, 1889, II.

<sup>3</sup> *Journal of Nervous and Mental Diseases*, 1890.

<sup>4</sup> *Berliner Klin. Wochenschrift*, 1890. <sup>5</sup> *University Medical Magazine*, 1890.

<sup>6</sup> *Neurologisches Centralblatt*, 1891. <sup>7</sup> *Ibid.* <sup>8</sup> *Ibid.* <sup>9</sup> *Ibid.*

<sup>10</sup> *Medical Record*. New York, March 12th, 1892. <sup>11</sup> *Medical Chronicle*, 1892.

<sup>12</sup> *Neurologisches Centralblatt*, 1892. <sup>13</sup> *Ibid.*

<sup>14</sup> *Zeitschrift für Klin. Medicin*, Bd. XX.

<sup>15</sup> *Journal of Nervous and Mental Diseases*, 1892.

<sup>16</sup> Quoted in *Neurologisches Centralblatt*, 1893.

<sup>17</sup> Quoted in *Revue Neurologique*, 1893. <sup>18</sup> *Neurologisches Centralblatt*, 1893.

<sup>19</sup> *Archiv für Psychiatrie*, Bd. XXV.

<sup>20</sup> *Journal of Mental Science*, Vols. XXXVIII. and XXXIX.

<sup>21</sup> These have already been published in a paper "On Chronic Chorea," *Journal of Nervous and Mental Diseases*, 1893.



His swaying, jerking, and fantastically irregular walk compelled him from the sidewalk to the unobstructed roadway. Notwithstanding his infirmity, he was a great pedestrian, frequently walking from his home, eight miles distant, and returning the same day. His sudden stops and precipitate advance, his facial contortions and mobile features, I recall with great vividness after forty years." His wife died in childbed.

Margaret married J. M. Her symptoms began to develop before she was 40. She continued to go about until a few days before her death, which occurred in her 65th year. Except a short time before her death, she was not entirely helpless, nor were the mental symptoms very strongly marked in her case.

#### *Second Generation.*

Margaret M., the last-mentioned patient, had five children, two of whom have already died of the disease, and three are in various stages of it. I have seen two members of the family, and have performed a post-mortem on a third:—

*First child*, male, now in his 61st year. A year ago the first evidences began. "A man of some character, it is but charity to ascribe the eccentricities of his life to disturbed mentality. He married twice, but had issue only by his first wife. Several children died in infancy, but one surviving is now in good health." This patient I could not see.

*Second child*, female, married, became choreic in her 40th year, and died demented in December, 1890, in her 58th year. She was confined to her bed for nearly a year before her death, which occurred in the Pennsylvania Hospital for the Insane, Norristown. She had four children—three girls and one boy; all are living and in good health, the oldest being now in her 32nd year.

*Third child*, male, aged 55. I saw this patient with Dr. Ellis. He has enjoyed good health, and has been able to attend to his business until recently. When about 42 he began to get nervous. Irregular locomotion was the first symptom; his speech became affected about a year ago. He will make use of a nod or a grunt in place of words whenever he can. Lately he has been confined to the house, and has been obliged to abandon business. He is very irritable, and is steadily passing into a state of dementia. He has had five children: four are living and in good health, the oldest about 33 years of age; one died of basilar meningitis at 16. I saw this patient in April, 1889, and made the following note:—



Bony, well-built man ; face has an intelligent expression. The gait is very peculiar ; he sways from side to side ; the movements are irregular, very unlike those of an ataxic, but resemble rather those of an alcoholic. He does not use a cane ; in walking the feet are not specially spread ; the eyes are not directed to the ground. He can stand with his heels together, and with his eyes shut ; there are no movements of the hands or arms when at rest, but in attempting to move there are large irregular sweeps of the arms, and slight tremor. He has great difficulty in feeding himself, and sometimes takes two hours or more at a meal. He still can write, though with increasing difficulty. He can sign his name, but the pen, in forming the letters, is often jerked up and the signature is very irregular. With the eyes shut he touches the nose or ear with precision and quickly. The grasp of the hand is firm and strong. There is no disturbance of sensation, no numbness or tingling. The knee-jerks are slightly increased ; the ankle clonus is not obtainable. The pupils are of medium size, and react to light and on accommodation. The speech is slow, and interrupted frequently by the interjection 'Hem, ha !' This peculiarity, his wife says, is of comparatively recent development. The mental condition is apparently good ; perception clear. When questioned, however, on several occasions, it seemed to take him some time to understand our wishes. He takes an interest in what is going on ; reads a good deal, particularly the newspapers.

Within the years which have elapsed since making this note he has steadily declined mentally and bodily.

*Fourth child*, female, aged 43, married, has had five children. One died of scarlet fever ; the others are living, the oldest a boy of 23. In this case the disease has progressed with greater rapidity than in the others, and certain indications of it have been present, according to the doctor, since her thirty-fourth year. The mental symptoms were first to appear. In April, 1889, I made the following note :—

Slightly built, somewhat anæmic woman ; she talks clearly and rapidly, but occasionally displays a certain childishness, and the doctor, who has not seen her for some years, was much struck with the change in this respect.

While sitting quietly there were no irregular movements of her limbs, but occasionally there was a slight jerk of the finger, the shoulders would move, and once or twice, while speaking, there appeared to be irregular contraction of the facial muscles. There



is no tremor of the tongue, and the pharyngeal muscles act normally; the grasp is good; she can use her fingers for delicate movements, and can thread a needle, and there does not appear to be the slightest inco-ordination. The most marked change appeared to be noticed in her gait. She walks with the feet somewhat spread, and sways, though she follows a straight line fairly well; she turns with a little difficulty, and, if rapidly, loses her balance. Her head is carried somewhat stiffly in walking; she does not trip, and she walks in the dark quite well. She stands with the eyes shut and the feet together without swaying.

The muscular power of the legs is good; the knee-jerks are increased on both sides; there are no disturbances of sensation; the special senses are normal; the pupils are of medium size and react to light and on accommodation. In the years since the preceding note was made she has lost ground rapidly, and the muscular inco-ordination has become much worse. She is now confined to the house, and for the greater part of the time to her bed.

*Fifth child*, female, aged, at the time of her death, 51; married; had eight children. Dr. Ellis writes: "After the birth of the seventh child, in her thirty-second year, her husband noticed the beginning of the trouble in jerking movements of the legs when sitting, and when erect she had a trick of raising her heels suddenly and standing upon the balls of the toes. Irregular movements of the arms speedily followed. When I saw her first, in 1880, she could walk a mile or two without apparent fatigue, and would insist on walking to church, nearly a mile distant, repelling the suggestion that she could not walk as well as another. At this time, in walking, her body would be bent forward, her head jerking, with a pendulum-like motion, to and fro, and her legs making such irregular and large movements that she would make wide excursions on the sidewalk. A year later she could no longer go out without assistance. Her speech indicated marked changes very early, in her fortieth year, and this was (in 1881 and 1882) accompanied by great difficulty in swallowing and frequently with alarming spells of strangling. She was a most pitiable sight. She suffered also from procidentia uteri; yet in June, 1883, in her forty-third year, she was delivered of her eighth child, which survived but a few days. Her menses were perfectly regular, the menopause not occurring until the forty-eighth year. Six months before her death she was confined to her bed, utterly helpless, and was fed with a spoon. She was now entirely demented.



"The deep reflexes were rather exaggerated. She could go about the house at night with as little help as in the daylight. She was exceedingly irritable and cross. The choreic movements stopped in sleep; there was no palsy of the sphincters. Of her eight children, seven are living, the oldest in her thirty-third year; all are in good health."

*Post-mortem* (about thirty hours after death).—Considerable wasting of the body; no enlargements of joints; no abnormal position of limbs; face a great deal wasted, presenting several recent scars and abrasions, the result of falls.

The skull-cap was of moderate thickness; the dura was tense; the meningeal vessels looked stiff; the longitudinal sinus contained recent clots. On the exposed cortex cerebri the arachnoid was somewhat turbid, and universally separated from the pia by a considerable amount of serous exudate; this was especially marked over the sulci. The Pacchionian granulations were numerous; the cortical veins moderately full. At the base the arachnoid was turbid, and the larger arteries a little stiff; the meninges were not specially adherent, and the pia could be stripped without tearing the substance. Superficial examination revealed no areas of softening, and no special lesions of hemispheres or of cerebellum. There was general wasting of the convolutions, which were also, on section, rather firm. The gray matter was dark, and in places looked thinner than normal. The crura presented no signs of descending degeneration; the pons and medulla were natural-looking; anterior pyramids had a clear, normal aspect; the ventricles were not distended. The spinal cord was firm; arachnoid a little opaque; pia normal. Transverse sections showed no systemic degenerations; the gray matter had a rosy red tint.

*Microscopical Examination.*—I am indebted to Dr. Gray for an extensive series of sections from various parts of the brain and cord. The changes may thus be summarized: The arteries were thickened and in places showed hyaline degeneration; in the smaller arterioles fatty changes were very marked in the fresh specimens from the cortex. Here and there the perivascular lymph-spaces were large and contained leucocytes. The ganglion cells in many sections showed very slight changes, not more than are often seen in chronic disorders associated with atrophy of the convolutions. There was the common vacuolation, and many cells seemed laden with pigment. The increase in the connective-tissue elements was more evident to the touch and on section than microscopically. Sections



of the pons and medulla showed no special foci of disease. Beyond thickening of the arteries and a shrinkage in the cells of the anterior cornua (probably an artificial change), the sections of the cord showed no lesions.

*Neiter Family.*

So far as can be ascertained only four members of the family have been affected, namely, mother and three children, one of whom was our patient, Peter.

1. The mother, a German, is stated to have had trouble of the same kind as that which Peter has. For many years she made wild inco-ordinate movements with her arms, and toward the end of her life she could not eat alone and had to be fed. Her mind, also, became very weak. The exact duration of the disease in her case could not be obtained, but it extended over many years. She is said to have died of heart disease. She has one brother living, aged 83, who is said to have the disease, but Dr. Chas. Simon, who visited him, reports that he is the subject of ordinary senile tremor. No information is available with reference to her family. Her maiden name was Schmidt. She had four children, of whom three have been affected with the disease.

2. Lizzie N. was well to her 37th year. Married and had six children, of whom two died and four are living and well. After the birth of the last child, the chorea developed, beginning first in the arms. Her husband noticed that she frequently dropped things. The trouble gradually became worse. Her mind became seriously affected, she talked incoherently, and had strange ideas. She once tried to commit suicide by jumping out of a window. During the last year of her life she was helpless, and could not walk alone. She died in her 49th year, about twelve years after the first onset of the symptoms. Her husband, from whom these facts were obtained, says that the disease was called St. Vitus' dance.

3. Nicholas Neiter, aged about 40, blacksmith, living at Edgewood, Hartford Co., Md. He was seen for me by Dr. Chas. Simon, who reports that he is evidently subject to the disease, as he displays grotesque inco-ordinate movements of the legs, arms, and face. Mentally, too, he is inclined to be childish and is very emotional. He regards himself, however, as in a condition of perfect health and not affected in any way as his brother Peter.

4. Peter Neiter, aged 59, German, a butcher, was admitted to



Johns Hopkins Hospital,<sup>1</sup> October 9, 1890. Patient has been in this country since 1850. He has always enjoyed good health with the exception of malaria when he first came to this country; he has not had syphilis. He dates the present trouble from an attack of gastro-intestinal disturbance eight years ago, which followed the drinking of large quantities of iced lemonade. At this time he had also pains in the head, and he speaks of the occurrence of something bursting in his body like a cannon. The movements began about five days after this over-heating and taking iced drinks. They did not start at any particular part of the body, but were general from the outset. They have gradually become worse, particularly when voluntary movements are made. They are severe enough to prevent him from working, and he has not been able to do much for six or eight years. He has fallen sometimes, owing to the irregular movements of the legs. He has never at any time lost consciousness. Emotion or fright always exaggerates the movements. He has not had headaches, and, as a rule, sleeps well. His appetite has been good and general health excellent. Ever since the attack, eight years ago, he has been liable to a recurrence of the vomiting whenever he takes cold drinks. He says his memory is quite good. He does not think that his speech has been affected.

*Present Condition.*—The patient is a large, well-nourished, well-built man. The face in repose looks intelligent, but on smiling, the expression is fatuous. He answers all questions readily and freely; gives a good account of his condition, and it is more in his expression and general behaviour that an indication is found of mental impairment.

When sitting in a chair, at ease, the arms and hands are in more or less constant irregular motion. The fingers are extended and flexed alternately; sometimes only one, sometimes the entire set. At other times the whole hand will be lifted, or there are constant movements of pronation and supination. For half a minute or so they may be perfectly motionless. The head and trunk present occasional slow movements; in the latter more of a swaying character. The legs jerk irregularly and the feet are flexed or extended; but the movements are not so frequent as in the arms. The face in repose is usually motionless, but the lips are occasionally brought together more tightly and the chin elevated or depressed. There is an occasional movement of the zygomatic and of the

<sup>1</sup> The patient was shown at the Hospital Medical Society, and is reported in the Hospital Bulletin, Vol. I.



frontal muscles. He puts out the tongue, with tolerably active associated movements of the face, and it is usually quickly withdrawn or rolled from side to side. It is impossible for him to hold it out for any length of time. There are no irregular movements of the palate muscles.

He walks with a curious irregular gait, displaying distinct inco-ordination, swaying as he goes, hesitating a moment in a step, keeping the arms out from the body and in constant motion. The legs are spread wide apart; the steps are unequal in length and he seems rather to drag the feet. He stands well with the heels close together and the eyes shut.

There is a suggestion of stiffness about the gait and about the way in which he uses his legs.

The sensation is unaffected. The deep reflexes are increased. There is slight ankle clonus, the knee-jerks are exaggerated, and there is slight increase in the arm-reflexes.

The special senses are unimpaired. The pupils are of medium size—the right a little larger than the left; they react to light and on accommodation; there is no nystagmus. He has no fever; the bowels are regular, and the urine shows no special changes.

**Special Features.**—The *onset* is gradual in the hereditary form; but there are instances, as Observation XI. in Huet's monograph and Berkeley's case,<sup>1</sup> which followed emotional causes and came on suddenly.

The earliest manifestations are motor. It is first noticed that the gait is not quite so steady as usual, or that there are slight irregular movements of the hands. In a few cases the mental disturbance has been the first symptom, but, as a rule, there is no perceptible disturbance in the intellectual faculties until the motor features of the disease are well developed. In the three cases which I saw in the Family X the disturbance in locomotion was the first symptom observed.

*Character of the Movements.*—Though irregular, involuntary and arrhythmic, the movements in the chronic progressive form differ in one important particular from those of Sydenham's chorea, namely, in the absence of the brusque, quick, jerking character. At first indeed the condition is one rather of muscular instability or inquietude, and when the patient is at rest there are irregular movements of the muscles of the hands or arms, which perhaps scarcely alter the position of the limb, or slight, slow contractions

<sup>1</sup> *Medical News*, 1883, Vol. XLIII.



pass over the muscles of the face. There may be no movements of the hands or arms when in repose, but any attempt at grasping an object may be associated with large, irregular, sweeping movements, as were present in a very striking manner in the third child in the second generation of Family X. In this case and in his sister the movements of the arms were rather those of inco-ordination than truly choreiform, and yet such a voluntary effort as the threading of a needle could be performed by the latter. In fact the two points of difference in the movements from those of Sydenham's chorea are the slowness and the inco-ordination. The slowness of movement is noticed particularly in the facial grimaces, which I have never seen presenting that rapidity so characteristic of chorea minor. These differences have not always been appreciated by writers. Thus E. S. Reynolds and Menzies state that the movements are exactly like those of ordinary chorea. Doubtless, the cases differ somewhat in this respect, but in those which have come under my observation the points of contrast above referred to were very manifest.

One of the most striking peculiarities is the gait, which, as in the members of Family X, may be very early affected. It has been best described by King, who states very correctly that when fully developed it constitutes one of the most important features of the disease, and is unlike the gait in any other affection. The station may be good with the exception of a slight swaying of the trunk, but on attempting to walk the unsteadiness develops and is characterized by large lateral deviations from the straight line, by marked swaying of the body and sometimes by precipitate movements, in which the patient almost falls but catches himself. As mentioned by Dr. Ellis, even when this gait is well developed the patient may be able to take long walks. It has been very well compared with the gait of a drunken man. The difficulty in locomotion may persist for many years before the patient becomes bed-ridden.

In a majority of the cases *speech* is affected, at first manifested in a slowness and hesitancy, and in the interjection of such expressions as hem! ha! &c., and finally by great indistinctness, owing to inability to pronounce the words clearly. It has not the staccato, scanning quality of the speech of multiple sclerosis.

The handwriting is early affected; the letters are badly formed, irregular, and run into each other, and the lines are not straight, but with large zig-zags. In the later stages writing becomes impossible.



Under the influence of emotion, as during examination, the movements are usually aggravated. The influence of the will may control them to some extent in the earlier periods of the disease, but, as a rule, when the disease is well developed, the movements are not influenced by any voluntary effort of the patient. One member of the Family X, in whom the disease was quite evident, could thread her needle without difficulty.

*Sensation* is not affected, nor are the special senses involved. The muscular force is retained until the disease is well advanced. The reflexes are usually increased, and the knee-jerks may be exaggerated at an early stage, as in the fourth child in Family X.

Huntington gave as one of the three special features of the disease "a tendency to *insanity* and suicide." This has been confirmed by subsequent observers. At first there may be only irritability or moodiness and depression, but the most constant change is a slow but progressive enfeeblement of the mental faculties, without, as a rule, hallucinations, or delusions of grandeur or of persecution. The disposition to suicide has also been observed by many writers since Huntington.

*Heredity* is one of the most remarkable features of the disease; indeed, in the whole range of inherited disorders there is scarcely one in which a larger percentage of individuals have been found affected. There are families in which 25 per cent. of all the members known have been attacked, and more than 50 per cent. of adults. The tendency appears to be transmitted through male and female alike, and the sexes are about equally attacked, though in some families the males have suffered most severely. Isolated cases occur in members of perfectly healthy families, so that the designation chronic progressive chorea, which has been recommended, is more correct than that usually employed, and which lays the chief stress upon heredity. The *age* of onset, the third feature which Huntington regarded as characteristic, is usually, but not always, in middle life. In a very large proportion of the cases with an hereditary taint, the disease does not begin until after the 30th year. Huet gives seven instances of this form, in which the disease began before this date. In Hoffmann's patient the affection began with adolescence.

**Morbid Anatomy and Pathology.**—Only a few cases of the hereditary form have been carefully studied. In Huber's case, in which the disease had lasted for eight years, there was pachymeningitis and chronic adhesions between the pia and the cortex.



In the sisters described by MacLeod,<sup>1</sup> in one case which had lasted about six years there was pachymeningitis with hæmatoma on the left side, and atrophy of the convolutions; the pia mater was adherent, and the ventricles were somewhat dilated. In the other case the chorea had come on late in life, about the 70th year, and death had followed in the 73rd year. The autopsy showed fibrous tumours of the dura mater on the left side, chiefly in the neighbourhood of the bases of the first and second frontal convolutions. In a case examined by Klebs,<sup>2</sup> in addition to hæmorrhagic pachymeningitis, he found foci of cell infiltration in the white matter, and hyaline thrombi in the vessels.

Greppin<sup>3</sup> has contributed a very careful study of a case from Professor Wille's clinic at Basel. There was a moderate grade of atrophy of the convolutions, and microscopically a condition of chronic encephalitis, consisting chiefly of small and large foci of cell infiltration, particularly in the superficial white matter in the frontal and temporal lobes.

Kronthal and Kalisher's<sup>4</sup> study of a case which had lasted for fifteen years is particularly thorough. They found chronic pia-arachnitis with adhesions of the membranes to the cerebral cortex, particularly in the frontal region and over the central gyri; thickening of the vessel walls and small celled infiltration about them; diffuse degeneration of the pyramidal tracts of the pons, medulla, and antero-lateral columns of the cord, with other changes of less moment. The authors were not prepared to assert that any of these conditions were connected causally with the chronic progressive chorea.

In the monograph of Huet, eight autopsies are given in cases of chronic chorea without hereditary basis. In the case reported by Macleod, the man had had chorea for six years with dementia. There were pachymeningitis and hæmatoma of the dura mater on both sides. In Maclaren's case, the man, aged 38, had had chorea for eight years and progressive dementia. There was thickening of the membranes with atrophy of the convolutions. In Berkeley's case, a man, aged 41, had had chorea for seven years and gradual dementia. There was atrophy of the dura, and microscopically dilatation of the arteries with thickening of the walls, small areas of softening, and vacuolation of the nerve cells. In the case reported by Bacon, the woman, aged 61 had had chorea for three years.

<sup>1</sup> *Journal of Mental Science*, 1881.

<sup>2</sup> *Correspondenzblatt f. Schweizer Aerzte*, 1888.

<sup>3</sup> *Loc. cit.*

<sup>4</sup> *Loc. cit.*



There was atrophy of the cortex with a chronic arachnitis. In the case of Vassitch, a woman, aged 41, had had chorea for seven years with gradual mental impairment, and delusions of persecution. She had previously had hysteria. No lesions were found at the autopsy. In the case of Pau, a man aged 55 was admitted to the asylum, and died in six months. There was atrophy of the cortex with increased density. In Tissier's case, a woman, aged 79, had had chorea for two years with mental enfeeblement. The pia mater was a little thickened, was only adherent where there were some spots of cortical softening, which were chiefly in the external portions of the occipital lobes, and the second and third frontal convolutions of the left hemisphere.

The most recent contribution is by Oppenheim and Hoppe,<sup>1</sup> who have studied two cases; the first a woman, aged 55, in whom the disease had lasted from her thirty-second year. There was hydrocephalus externus, and turbidity of the membranes; the convolutions were atrophied and the sulci deep. The ventricles were not enlarged. A disseminated encephalitis was manifest by the presence of foci of small round cells, chiefly in the central convolutions, and there was atrophy of the layer of small round cells in the grey cortex. There was some diffuse sclerosis also in the antero-lateral columns of the cord. In the second case, which died at the age of 75, the disease began late in the seventieth year. A chronic pachymeningitis hæmorrhagica was found with moderate atrophy of the convolutions, and in the region of the central convolutions there were foci of miliary infiltration and atrophy also of the small round cells in the border between the first and second cortical layers. From their investigation, with which those of Golgi, Klebs, and Greppin practically agree, they regard the disease anatomically as a cortical and sub-cortical miliary disseminated encephalitis.

Menzies<sup>2</sup> describes one autopsy in which he found atrophy of the cortex cerebri, with increased consistence, and microscopically coarse neuroglia, thickened vessels and degeneration of the nerve cells of all the layers. In brief the changes belong to those degenerative processes in the grey cortex which are common to so many affections associated with impaired motor and psychical functions.

So far nothing has been found which is peculiar to the disease or in any way specific. Of the manner in which these changes are initiated, of the nature of the first slight departure from the normal, of the character of the differences in the morbid processes which

<sup>1</sup> *Loc. cit.*

<sup>2</sup> *Loc. cit.*



make the external picture that of chronic progressive chorea and not that of, say, double athetosis, why in the members of certain families, after a certain age, the malady is liable to occur, why other individuals cast in the same family mould should escape—of these and other questions science at present offers no solution.

**Diagnosis.**—The recognition of chronic progressive chorea, so easy in the family form, becomes exceedingly difficult in cases without this peculiarity. There can be no question, in the first place, that cases with and cases without hereditary features present identical symptoms and course. The term may be used to group the cases which have a chronic and progressive course. The heredity, as in Friedrich's ataxia, is only one, and an inconstant feature of the disorder. The "progress" is towards motor and mental enfeeblement.

The cases of chronic chorea dating from birth, or shortly after, almost invariably have *spasm* as an associated feature, a symptom which excludes a case, I believe, from the category under consideration. In the later stages, after many years, there may be increased reflexes, and some rigidity in chronic progressive chorea, but it does not form a feature in the fully-developed disease.

The following are good illustrative cases of the so-called congenital and spastic chorea<sup>1</sup> :—

*Case 16.*—Mary M——, æt. 4. Breech presentation, delay at the head; was six hours before she was resuscitated. Began to talk at two years; never walked; almost from birth she has had peculiar movements of hands and arms; the thumbs are turned in and there is constant irregular motion of the arms and hands, with stiffness, which is made worse when she attempts to control it; it is like a chorea. There is also some co-ordination of the head. She is well nourished; no wasting. Co-ordination of legs good; but she does not walk.

*Case 17.*—Nellie P——, æt. 9. Parents healthy, five children dead of seven. Seven years ago had fits while teething, had fits constantly for twenty-one days; for nine months had seven to nine per diem; in very weak health when fits ceased. *Present state:* Speech hesitating; memory not affected; unable to stand, sit, feed herself, or assist herself in any way; can move every muscle in the body, but with an irregular movement which prevents her using any group of muscles; the movement is choreoid; in attempting to grasp an object the fingers are thrown out in a stiff,

<sup>1</sup> From my "Cerebral Palsies of Children," p. 66.



spasmodic, and irregular manner, and she is unable to close them over the object.

So also from double athetosis, certain forms of which resemble closely chronic progressive chorea, the element of spasm is the most important differential sign. As Audry, in his admirable monograph on Double Athetosis,<sup>1</sup> says:—The athetoid movements in the limbs are associated with rigidity, even with contracture of the muscles. It is a symptom essentially spasmodic, the reflexes are found exaggerated when the spasm of the muscles is sufficiently easy to permit of the trial. The gait also is spastic.

From the various forms of chronic generalized tic there could rarely be any difficulty, the character of the movements are so radically different. Charcot, however, gives a case<sup>2</sup> of tic which simulated somewhat Huntington's chorea.

With the exception of Charcot, and his pupil, Huet, all the writers on chronic progressive chorea regard the disease as totally different from chorea minor, a view which seems to me just, when we take into consideration the clinical features, particularly the character of the movements, the progressive course, the heredity, and the anatomical lesions.

Unhappily the *treatment* of chronic progressive chorea is futile. In the words of the great physician, whose loss all clinical physicians deplore, "son évolution est fatale, et la thérapeutique est impuissante à l'arrêter, ne fut-ce qu'un instant, dans sa marche progressive."

<sup>1</sup> *L'Athétose Double*. Paris, 1892.

<sup>2</sup> *Leçons du Mardi*, 1888-89, p. 463.



# APPENDIX.

## ANALYSIS OF SEVENTY-THREE FATAL CASES OF CHOREA MINOR.

No.	Sex.	Age.	Rheumatism.	Endocarditis.	Pericarditis and other lesions.	Remarks.	Reporter.
1	M.	17	Rh.	Mitral	—	First attack	S. Mackenzie, <i>Trans. International Medical Congress</i> , 1881, Vol. iv. p. 99
2	F.	19	No rh.	Mitral	—	Second attack. Pregnancy	Ditto
3	F.	10	No rh.	Aortic and mitral. Mitral stenosis	—	Second attack	Ditto
4	F.	19	(?)	Fatty heart	—	Pregnancy; abortion. Pyæmia	Ditto
5	F.	27	No rh.	Mitral	—	Pregnancy	Ditto
6	M.	13	Rh. subacute	Mitral and tricuspid	Pericarditis	Death from pericarditis	Ditto
7	F.	13	No rh.	Mitral, aortic and tricuspid	Pericarditis, pneumonia and pleurisy	About a month's illness	Bouchut, <i>Paris Médical</i> , 1881, vi., 129
8	F.	8	No rh.	Mitral	Infarcts in kidney and spleen	Second attack, T. 101°. Death from cerebral hæmorrhage	Baxter, <i>Brain</i> , April, 1879
9	F.	20	No rh.	Mitral	—	First attack, shock and excitement, very severe chorea insaniens, T. 39.6°. Acute onset	Frank, <i>Allg. Wiener Med. Zeitung</i> , 1879



No.	Sex.	Age.	Rheumatism.	Endocarditis.	Pericarditis and other lesions.	Remarks.	Reporter.
10	M.	18	Ac. rh.; chorea followed	Mitral	Spleen enlarged	Wetting. First attack, T. to 38°2.	Maixner, <i>Med. Chir. Centralblatt. Wien</i> , 1882. Bd. xvii.
11	F.	20	No rh.	Heart normal, but well-marked murmur during life	—	Fright. Chorea insaniens. T. 107°. Death within a week	Donkin and Hebb, <i>Med. Times and Gazette</i> , 1884, II.
12	F.	11	Rh. pains	Mitral and aortic endocarditis	Old pericarditis. Pleurisy	Third attack, T. 40°6°. Hæmaturia; skin eruption	Morrel-Lavellée <i>Revue des Maladies de l'Enfance</i> , 1884
13	F.	10	At five chorea and rheumatism	Aortic and mitral incompetency; mitral, tricuspid and aortic endocarditis	—	R. pains at onset of second attack. Death from chronic valve disease	Oxley, <i>Lancet</i> , 1886, Vol. II., p. 142
14	F.	19	Acute rh. six months before	Mitral endocarditis	Broncho-pneumonia	Second attack, Skin eruption, T. 39-40°1°. No murmur. Phlegmon	Guinon, <i>La France Médicale</i> , 1886, I., p. 73
15	F.	9	No rh.	Mitral	—	First attack. Very rapid case. Delirium, T. 101°. Death in 130 hours	Cook and Beale, <i>British Medical Journal</i> , 1888, I., 795
16	M.	18	Mental shock and cold, no rh.	Mitral	Hæmatoma of dura mater, hyperæmia of brain	T. to 41°5°. Sixteen days' duration	Grosse, <i>Berliner Klin. Wochenschrift</i> , Nos. 33 and 34, 1889



17	M.	66	No rh.	Mitral	—	Second attack, first six or eight years ago. Deep coma after bromide and chloral, four days after admission	Ditto
18	F.	21	No rh. Four previous attacks of chorea	Mitral	—	Chorea insaniens, T. to 107.6°. Duration about eleven days	Mitchinson, <i>Lancet</i> , 1889, I.
19	M.	16	Rh. acute, eight weeks before	Mitral	—	T. to 105°; delirium	Ditto
20	M.	19	"Some rheumatic symptoms"	Mitral	Lymph on arachnoid and on cortex cerebri	First attack. Acute chorea insaniens; admitted to asylum	Evan Powell, <i>Lancet</i> , 1889, I.
21	F.	20	No rh. Mental worry	Mitral cusps thickened, no vegetations	—	First attack. Chorea insaniens; admitted to asylum. T. to 102.5°	Ditto
22	M.	17	No rh.	Heart normal	—	Very severe and rapidly fatal case	Schrötter, <i>Wiener Med. Wochenschrift</i> , 1889, No. 18
23	F.	7	Rh. ac. previously	—	Pericarditis	T. 101.5°. Rapid case. Death from pericarditis	Cheadle, "Rheumatic State in Childhood," London, 1889, p. 81
24	F.	15	No history of rh.	Mitral	Pericarditis	Illness of three weeks. Violent chorea	Ditto p. 82
25	F.	20	Rh. chorea during convalescence	Mitral	—	First attack. Fever, acute dilatation of stomach	H. H. Brown, <i>Lancet</i> , 1890, II.



No.	Sex.	Age.	Rheumatism.	Endocarditis.	Pericarditis and other lesions.	Remarks.	Reporter.
26	F.	12	Rh. ac. two years before	Mitral insufficiency ; fresh mitral endocarditis	—	Second attack of chorea	Goodall, <i>Guy's Hospital Reports</i> , 1890
27	F.	19	Rh. ac. three months before, and on admission	Mitral and aortic	—	Thrombosis of right femoral vein, broncho-pneumonia	Ditto
28	M.	8	Ac. rh.	Mitral incompetency ; fresh mitral and aortic endocarditis	Pericardial adhesion	Second attack of chorea	Ditto
29	F.	11	Rh. slight	Mitral	Pericarditis. Phlebitis, Empyema	Second attack	Ditto
30	M.	19	Ac. rh. two months before	Mitral	—	Second attack. Mania	Ditto
31	F.	17	No rh.	Mitral and aortic	Pericarditis, acute nephritis	Second attack	Ditto
32	M.	18	Rh. ac.	Mitral stenosis. Ulcerative endocarditis of mitral and tricuspid valves	Infarcts ; gangrene of foot	Frequent attacks	Ditto
33	F.	27	Ac. rh. four years before	Mitral incompetency ; fresh endocarditis	Infarcts	First attack	Ditto
34	F.	7	Rh. (?)	Aortic and mitral	—	Third attack	Ditto







No.	Sex.	Age.	Rheumatism.	Endocarditis.	Pericarditis and other lesions.	Remarks.	Reporter.
45	M.	16	No rh.	Heart normal	—	First attack	Pianese, <i>La Natura infettiva della Corea del Sydenham</i> , Napoli, 1893
46	M.	17	Rh. ac. ten weeks before chorea	Aortic and mitral endocarditis	No special change in brain or cord.	Maniacal chorea, T. to 105·6°. Death within ten days	Affleck, <i>Edinburgh Hosp. Reports</i> , Vol. I, 392
47	F.	14	No rh. After about five weeks' fever, then "rheumatoid pyæmia"	Mitral	Parotitis	T. to 103°. Choreia insaniens	Gee, <i>St. Bartholomew's Hosp. Reports</i> , xxii.
48	F.	9	Rh. (?)	Mitral	Intense hyperæmia of brain	Decubitus; coma	Koch, <i>Deutsches Archiv f. Klin. Med.</i> , Bd. xl.
49	F.	13	Rh.; chorea followed in one and a half weeks	Mitral	Pericarditis	Pleur. sin. Two weeks' illness	Ditto
50	M.	60	No rh.	Mitral	—	An attack of chorea 8 years before	Ditto
51	M.	18	No rh.	Mitral	Hæmatoma of dura mater	Followed excitement and cold. Very acute case, fatal in sixteen days	Ditto
52	M.	17	Rh.	Mitral	Acute pericarditis	T. to 39°	Wollenberg, <i>Archiv f. Psych. and Nervenkrankheiten</i> , Bd. XXIII.



	F.	II	(?)	Mitral, old and recent	—	—	Ditto
53							
54	F.	7	No rh.	Mitral and tricuspid	Slight pericarditis, acute	T. 103°-105°; acute choreic endocarditis	Jas. Finlayson, <i>Archives of Pediatrics</i> , vol. VII., p. 497
55	F.	15	Scarlet fever, arthritis, and then chorea	Old mitral insufficiency, mitral endocarditis	Pericarditis	Second attack, chorea at 7th year, pleurisy; chill, T. 41	Litten, <sup>1</sup> <i>Charité Annalen</i> , Bd. XI., p. 263
56	F.	(?)	Primipara, scarlatina, arthritis	No endocarditis	—	Severe post-partum chorea with septicaemia, diphtheritic endometritis; infarcts, &c.	Ditto
57	F.	23	Abortion third month, Polyarthritis suppurativa	Ulcerative, mitral	—	General chorea on tenth day, first attack; septicaemia	Ditto
58	F.	25	Chorea at 14, arthritis followed by chorea	Chronic endocarditis. Fresh warty endocarditis	—	Second attack; purpura; fever, T. 40·1°	Ditto
59	F.	17	Chorea in 14th year. Ac. rh. at 15 with relapses	Old aortic lesion, fresh aortic endocarditis	—	Cold; arthritis, fever, chorea	Ditto
60	F.	17	No rh.; hæmaturia, menæna	Mitral	Acute nephritis for a week	Peritonitis, pleurisy and pneumonia. Death in seventh week of illness.	Turner, <i>Trans. of the Pathological Society of London</i> , Vol. XLIII.
61	F.	12	Ac. rh., three months later chorea	Heart hypertrophied. Fresh mitral endocarditis	—	Pleurisy	Ditto

<sup>1</sup> I have not included two or three of the older cases given by Litten, as they possibly have been included in other statistics.



No.	Sex.	Age.	Rheumatism.	Endocarditis.	Pericarditis and other lesions.	Remarks.	Reporter.
62	F.	16	Sub. ac. rh.	Mitral	—	Severe chorea insanien- s.	Turner, <i>Trans. of the Pathological Society of London</i> , Vol. XLIII.
63	F.	16	No rh. fright	Mitral	Abscess in neck	Violent chorea insan- iens. Fever, T. 101°	Ditto
64	F.	11	No rh.	Mitral	—	Emboli in corpora stri- ata. Diphtheria towards close	v. Starck, <i>Archiv f. Kinderheilkunde</i> , Bd. XIII.
65	F.	17	No rh.	Mitral	—	Very rapid case	Naunyn, <i>Mitt. aus der Med. Klinik zu Kö- nigsberg</i> , 1888
66	F.	7	No rh. cause	Mitral and aortic	Pericarditis; pneu- monia	Severe chorea. Rapid pulse, 37.9° highest fever.	Neuwerk, <i>Ziegler's Beit- räge</i> , Bd. I.
67	F.	11	Ac. rh.	Mitral and aortic	Pneumonia	—	Osler, <i>American Jnl. Med. Sciences</i> , 1887
68	M.	11	No rh.	Ac. ulcerative endo- carditis	Infarcts in spleen, kid- ney and brain	—	Ditto, also Molson, <i>Canada Med. and Surg.</i> , vol. xiii.
69	F.	18	No rh.	Mitral	—	Very acute case with mania	Ross, <i>Canada Med. and Surg. Jnl.</i> , Vol. XI. Osler, <i>American Jnl. Med. Science</i> , 1887, i.



70	F.	27	No rh.	Mitral	Parotitis	Acute maniacal chorea	Peakeley, <i>Johns Hopkins Hospital Reports</i> , Vol. II.
71	F.	70	No rh. Emotional disturbance	Old mitral lesion	—	Death in an attack of cardiac dyspnoea	Personal observation
72	M.	18	Rh. followed the chorea	Mitral endocarditis	Congestion of brain	T. to 40·3°	Charcot, <i>Leçons du Mardi</i> , 1888-89, p. 103
73	F.	9½	Rh. ac. following measles	Endocarditis	Pericarditis; pneumonia; death from former	Areas of sub-cortical softening	Patella, quoted in Schmidt's <i>Jahrbücher</i> Bd. 221







# INDEX.

Arithmomania, 87  
 Athetosis, double, 112  
 Auditory hallucinations in tic, 84

Balancing, 89  
 Barking cough of puberty, 95  
 Blind, forms of tic in the, 89

Chorea, canine, 17, 63, 68  
 ——— chronic progressive, 96  
 ——— ——— diagnosis of, 111  
 ——— ——— gait in, 107  
 ——— ——— heredity in, 97, 108  
 ——— ——— handwriting in, 107  
 ——— ——— insanity in, 97, 108  
 ——— ——— morbid anatomy of, 108  
 ——— ——— pathology of, 110  
 ——— ——— speech in, 107  
 ——— ——— suicide in, 97, 108  
 ——— ——— symptoms of, 99, 106  
 ——— congenital, 2, 39, 111  
 ——— corpuscles, 62  
 ——— electric, 75  
 ——— Huntington's, 96  
 ——— insaniens, 22  
 ——— hysterical, 2, 18  
 ——— major, 2  
 Chorée malleatoire, 19  
 Chorea minor, 3  
 ——— ——— acute forms of, 21, 22, 39  
 ——— ——— in adults, 44  
 ——— ——— in the aged, 44  
 ——— ——— age incidence of, 6  
 ——— ——— anæmia and, 17  
 ——— ——— apoplexy in, 61  
 ——— ——— arsenic, use of, in, 70  
 ——— ——— arthritis and, 13  
 ——— ——— atrophy of muscles in, 32  
 ——— ——— bacteriology of, 63

Chorea minor, blood in, 18, 60  
 ——— ——— baths, use of, in, 70, 71  
 ——— ——— cutaneous symptoms of, 35  
 ——— ——— chlorosis and, 18  
 ——— ——— definition of, 2  
 ——— ——— delirium in, 28, 35  
 ——— ——— diet in, 70  
 ——— ——— duration of, 39  
 ——— ——— embolic theory of, 61  
 ——— ——— endocarditis in, 30, 32, 33, 60  
 ——— ——— epilepsy and, 34  
 ——— ——— eye-strain and, 11  
 ——— ——— face, affection of muscles in, 29  
 ——— ——— ——— expression of, in, 35  
 ——— ——— family disposition in, 9  
 ——— ——— fatal cases of, 51, 60, 113  
 ——— ——— feeding in, 70  
 ——— ——— fever in, 37  
 ——— ——— fright as a cause of, 10  
 ——— ——— geographical distribution of, 9  
 ——— ——— hands, affection of, in, 29  
 ——— ——— hæmaturia in, 37  
 ——— ——— hæmorrhage as a factor in, 18  
 ——— ——— headache in, 20, 34  
 ——— ——— herpes zoster in, 35  
 ——— ——— hemiplegic weakness in, 31  
 ——— ——— heredity in, 10  
 ——— ——— heart in, 47  
 ——— ——— historical note on, 3  
 ——— ——— hysteria and, 18  
 ——— ——— infection, theory of, in, 64  
 ——— ——— imitation in, 10  
 ——— ——— infectious diseases and, 16  
 ——— ——— incontinence of urine in, 30  
 ——— ——— insanity in, 35  
 ——— ——— joints in, *see* Arthritis.



- Chorea minor, kidney, affections of, in,  
37  
— — — limbs, order of attack in, 29  
— — — locality, influence of, in, 9  
— — — massage in, 70  
— — — mental symptoms of, 34  
— — — micro-organisms in, 63  
— — — mild form of, 20  
— — — morbid anatomy of, 60  
— — — muscles in, 32  
— — — muscular weakness in, 31  
— — — naso-pharyngeal irritation in,  
11  
— — — nephritis in, 37  
— — — neuritis in, 33, 71  
— — — and old age, 44  
— — — optic neuritis in, 34  
— — — pains, in, 33  
— — — paralysis in, 31  
— — — pathology of, 64  
— — — pericarditis in, 58  
— — — pigmentation of skin in, 36,  
70  
— — — poisons as a cause of, 19  
— — — post-partum, 44  
— — — pregnancy and, 41  
— — — prognosis in, 39  
— — — psychical influences in, 10  
— — — purpura in, 36  
— — — race and, 7  
— — — recurrence of, 40  
— — — refraction errors and, 11, 12  
— — — rest, influence of, in, 69  
— — — retinal hyperæmia in, 34  
— — — retinal artery, embolism of,  
34  
— — — reflexes, state of, in, 34  
— — — reflex irritation in, 11  
— — — rheumatism and, 13, 67  
— — — school life, influence of, 10  
— — — scarlet fever and, 16  
— — — seasonal relations of, 5  
— — — sensation, disturbances of, 33  
— — — severe form of, 21  
— — — sex in, 6  
— — — site of changes in, 68  
— — — sleep, influence of, on, 30  
— — — spasmodic disorders, co-exist-  
ence with, 34  
— — — speech, disturbances of, in, 29  
— — — sub-cutaneous fibroid nodules  
in, 16  
— — — temperament and, 10
- Chorea minor, temperature, *see* Fever.  
— — — tic and, 40  
— — — traumatic influences in, 11  
— — — treatment of, 68  
— — — urine in, 37  
— — — uterus, irregular contractions  
of, in, 30  
— — — whooping-cough and, 17  
— — — mollis, 31  
— — — pseudo-, 2  
— — — rhythmic, 2  
— — — secondary, 2  
— — — spastica, 2, 39, 111  
— — — Sydenham's, 2  
— — — symptomatic, 2
- Choreiform affections, 2, 72
- Coprolalia, 87
- Cries, involuntary, 86
- Délire du toucher, 88
- "Dervish" tic, 90
- Dubini's disease, 75
- Echolalia, 79, 86
- Echokinesia, 87
- Electric chorea, 75
- Epilepsy and chorea minor, 34
- Epilepsia nutans, 89
- Expiratory cries, 90, 94
- Folie choreique, 35
- Folie pourquoi, 87
- Gait in chronic progressive chorea, 107
- Generalized tic, 75
- Gilles de la Tourette's disease, 79
- Habit spasm, 73
- Habit chorea, 73
- Head-nodding, 89
- Head-banging, 89
- Historical note, chorea minor, 3  
— — — chr. progressive chorea, 96
- Huntington's chorea, 96
- Hysteria and chorea minor, 18
- Hysterical cries, 91, 94
- Hysterical chorea, 2
- Ideas, fixed, 87
- Imperative ideas, 87
- Inspiratory cries, 90, 94
- Involuntary cries, 86



Jumpers of Maine, 87

Krouomania, 89

Latah, 87

Limp chorea, 31

Localized tic, 73

Maladie de la tic convulsif, 79

Mania, dancing, 4

Movements, character of, in chorea  
minor, 29

— — — in chr. pro. chorea, 106

— — — in tic, 84

Myriachit, 87

Obsessions mentales, 87

Onomatomania, 87

Pendulum spasm, 90

Ptussis canina, 95

Respiratory muscles, spasm of, 90

Simple tic, 73

St. Vitus' dance, 1, 3, 4

Tic, definitions of, 72

— forms of, 73

— with coprolalia, &c., 79

— generalized, 75

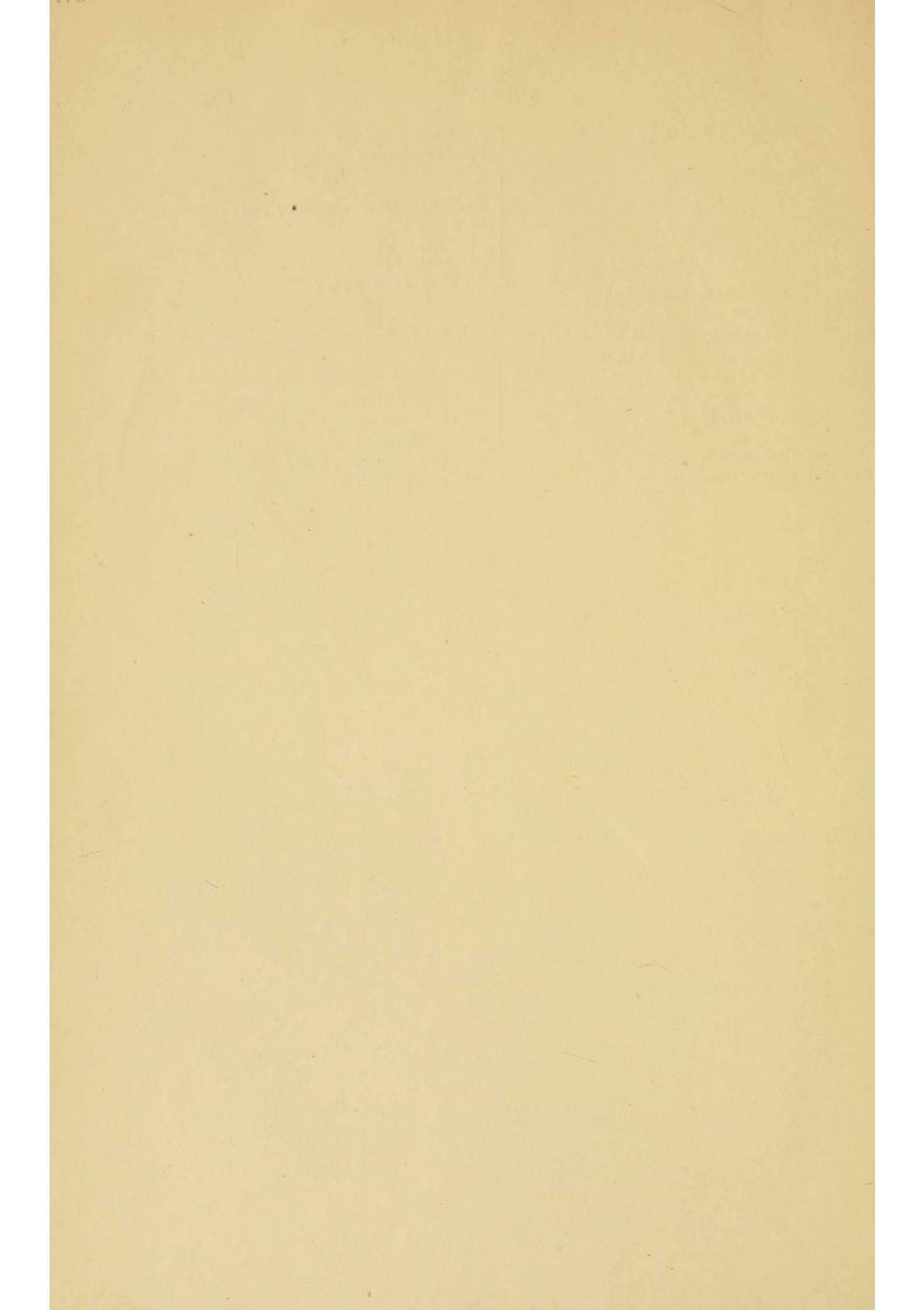
— localized, 73

— simple, 73

— complex co-ordinated forms, 88

— prognosis of, 88













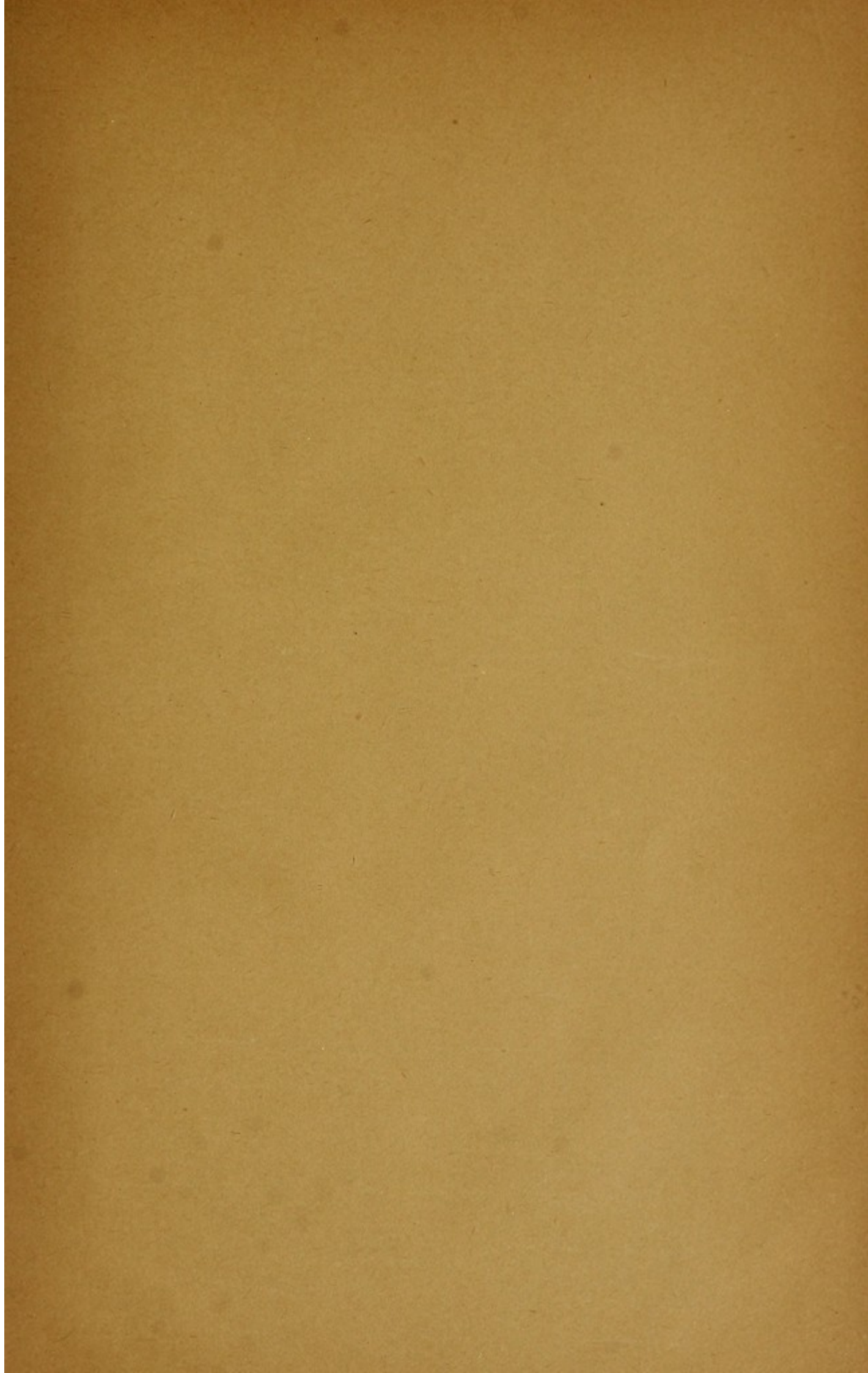














~~2/20~~  
134/10



