The choreas

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THE CHOREAS

A Senior Thesis prepared by

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There are many synonyms for the term Chorea among which are Infectious Chorea, Sydenham's Chorea, Sydenham's Disease, St. Vitus' Dance, Chorea Sancti Viti, Chorea of Childhood, Chorea Minor, Chorea Major, Dance de St. Guy, Periodical Factitation, Vit-Tanz and Chorea Germanorum. All of these names have been used with varied meanings in regard to their significance.

The word "Chorea" itself is derived from the Greek XOFEIA, meaning "to dance."

The Phrygian bacchantes, in their wild rites, were affected with violent autonomic movements, accompanied by disturbances of consciousness and the Suffi, in Persia, a sect which arose shortly after the founding of Mohammedanism, often passed into a condition of wild excitement, muscular spasms and general convulsions, during the dances connected with certain sacred ceremonies. This sect of the Suffi found many followers throughout Asia Minor, Persia, Egypt and Greece about the year 1000 A.D. At the time of the Crusades, the dance of St. John was an observed custom in Christian countries. (1) During an outbreak of a fresh religious excitement in Strasburg about 1418, the term "dance of St. Veit", first began to be freely applied to these disorders because those participating in the dancing mania were ordered by the chief magistrate to repair to the chapel of St. Vitus in Zabern, a village in Alsace. (1) The name St. Vitus seems to have it's origin from St. Veit, a young Sicilian boy who suffered martyrdom in the year 303 A.D. during the persecution of Diocletian. His body was carried from place to place by his followers and finally buried in the cloister of Korvey.

The name, St. Vitus Dance, is the only point in common between these
old religious dances and our present day "Idiopathic" Chorea.

Many German writers apply the term "Chorea Minor" to the affection in childhood, while they use the term "Chorea Major" or Chorea Germanorum" to designate affections resembling those of the hysterical epidemics of the Middle Ages or what is known today as "Hysteria Major."

(11) In surveying the literature of Chorea, I find many forms of so-called Chorea described. This multiplicity of forms is very largely due to the error of describing the symptom of choreiform movement as a disease or syndrome.

According to Tucker's classification (2) Chorea as an entity must conform to certain definite conditions, and all muscular movements disorders or symptoms not so conforming should be discarded. These criteria are:

1. Multiple muscular movements which must be spontaneous, irregular, involuntary and purposeless.

2. These movements must be reasonably traceable to the action upon certain portions of the brain of (a) a toxin from some form of infection, (b) some form of bacterial invasion, (c) some hereditary or acquired degenerative condition.

3. Chorea must be considered in the light of an encephalitis.

By adopting these criteria, we eliminate in our consideration of Chorea such phenomena as athetoid movements, the myoclonias, tics, habit or other spasms, torsions, rhythmic movements, drug poisonings causing muscular movements, functional movements, tremors and movements due to disease or injury which are not associated with encephalitis. Tucker (2) does not believe that Dubini's Electric Chorea with it's lightening like movements, convulsions, atrophy and reaction of degeneration, or that Bergerson's rhythmic movements occurring in anemic children should be
classified as Chorea.

Therefore, we shall confine our discussion of Chorea to the following seven forms and to Sydenham's Chorea in particular, as it is the form commonly seen by the general practitioner:

1. Sydenham's, or Acute Infectious Chorea.
2. Huntington's, or Chronic Family Chorea.
3. Hereditary, or Chronic Congenital Chorea.
4. Senile, or Arteriosclerotic Degenerative Chorea.
5. Chorea Gravidarum, or Chorea of Pregnancy.
7. Chorea of acute infectious diseases.

SYDENHAM'S CHOREA

Sydenham's or acute infectious Chorea may be defined as a disease occurring chiefly in children, due to some toxic or infectious agent, which acts on the central nervous system by producing irregular involuntary contractions of the muscles, resulting in purposeless movements, and associated with muscle weakness and mental irritability.

Frequency: - A great many mild cases of Chorea are accidentally discovered so that statistics are probably of no great value. Abt (3) found 226 cases of Chorea among 10,150 sick children. Osler reported 84 percent of a series of 500 and Starr listed 71 percent between 5 and 15 years.

Etiology: - There are many predisposing causes among which are age, sex, seasonal influences, race, heredity, social influence, emotional disturbance and others which will be considered. Statistics tend to show that most of the cases occur between the ages of 7 and 13 at the time when demands upon the nervous system are heavy and the metabolic rate is high.

Of 554 cases analyzed at the Philadelphia Infirmary for Nervous
Diseases, 71 percent were in females and 29 percent in males, according to Osler (5). The British Chorea Committee found in 436 cases, 114 males and 322 females (11). The proportion of males to females in all statistics is about 1 to 3.

All writers agree that most of their cases began in the spring and they attribute this to the fact that the general tone of the children is poor after being confined all winter and working hard at their school tasks.

Authorities do not agree as to the effects of climate on the disease. The prevalence is less in hot climates and the southern portion of the United States, except among negro children. In accordance with the well-known diathesis of Jews to nervous diseases, the incidence of Chorea is quite high in this race.

Children who live in tenements or under crowded, unsanitary conditions and suffer from malnutrition are more prone to contract the disease than those living under more favorable circumstances.

Strong emotional upsets and fright are very important predisposing causes but all authors agree that the interval between the fright and the onset of the disease must be from 3 to 5 days and not over a week if the fright is to be considered the predisposing cause (11).

Reflex irritation from the gastro-intestinal tract as in children infested with worms has been considered an etiological factor. Errors of diet, particularly a high meat diet is believed to play an important role in causing the disease and in these cases it has often been noted that the child was sleepless, had night terrors and enuresis before the attack.

Ritenour of Pennsylvania reports a case of Chorea in his own family caused by impacted teeth and which immediately cleared up following removal of five deciduous teeth which should have been cast off two or three years previously (6).
The direct cause of Chorea would appear to be, from the consensus opinion, some organism which becomes disseminated from some forms of infection as the teeth or tonsils. The role which rheumatic fever plays in the etiology is also important, in fact Chorea is usually mentioned as one of the cardinal symptoms of this disease.

Many men have endeavored to isolate the specific organism. Poynten and Paine cultivated the micrococcus rheumaticus in cases of articular rheumatism and found the same organism in the brain in cases of Chorea, associated with rheumatism (11). Helmholtz believes that the same organism which seem to tend to localize in the joints or the valves of the heart, may also localize in the cortex of the brain and produce Chorea (7).

In 1927 and in early 1928, J. C. Small (8) (9) published his articles on Streptococcus Cardioarthritis in Rheumatic Fever and Chorea. From a colored man whose illness began with tonsillitis with joint involvement several days later, he isolated gram-positive cocci in short chain formation to which he applied the above name.

Rosenow (10) isolated streptococci which grew on blood agar in small, dry, slightly elevated and non-adherent colonies surrounded by a greenish-brown zone, from the naso-pharynx and tonsils of patients having Chorea. After 24 hours a narrow zone of hemolysis developed, peripheral to the green zone. By injecting these bacteria into the roots of teeth of various animals, he was able to produce in rabbits a disease much resembling Chorea. In addition examination of the valves of the heart revealed a definite tendency of these organisms to localize there. He prepared a monovalent serum that had the property of agglutinating specifically the organism he has isolated.

The consensus of opinion seems to be that Chorea is a manifestation of rheumatic fever, in fact in many cases being the primary symptom antedating the joint involvement for weeks.

Diagnosis:— The diagnosis is made upon the history and physical find-
ings and by excluding those conditions resembling Chorea, but which by our classification, do not fall within the scope of true Chorea.

In children, the diagnosis as a rule is not difficult. Before the active symptoms are noticed, the parents will usually observe a clumsy attitude in the child especially at the dinner table. Over reaching and dropping the eating utensils are commonly reported. In a few days facial grimaces and aimless movements in one arm and shoulder make their appearance, and these purposeless movements, more marked while the child is observed, soon spread to the other side (12).

The most characteristic symptoms of the disease are rapid, coarse, involuntary, spontaneous, irregular and purposeless movements. These movements usually diminish during rest and cease during sleep, but are increased by excitement or restraint or the attempt to carry out volitional movements.

Weakness of muscle power is a common symptom and may be present before the choreic movements begin. If the patient is closely observed, it will be seen that one limb is used less than the other. The tendon reflexes are diminished or even absent and the muscle limp and hypotonic.

Laryngoscopic examination reveals an irregular movement of the vocal cords so that there is justification of including changes of pitch and intensity of the voice as one of the signs of Chorea.

Due to the involvement of the muscles of the eyeball, diplopia may be present. "Hippus," or spontaneous rapid and spasmodic variation in size of the pupil have been noted but the pupil is usually dilated, reacting to light and accommodation.

Fine tremor involving the fingers, tongue and lips may be also seen. This is probably due to the weakened general condition and is exaggerated by physical and mental excitement.

Cardiac symptoms are usually marked. The blood pressure is lowered due to the associated anemia and the pulse is increased in frequency 10-
15 beats per minute.

The endocarditis associated with Chorea is usually mild and may disappear, leaving no trace. Other cases leave permanent endocardial changes and valvular defects. Most of the symptoms referable to the heart are those due to mitral insufficiency.

Rheumatism is very commonly seen in Chorea, but should be considered a cause rather than a symptom.

Laboratory findings do not seem of much significance apart from an occasional positive blood culture or spinal fluid culture. A fairly constant low grade leucocytosis is usually found indicating the presence of infection. The urinary output is increased and crystals of urates and phosphates are abundant. A transitory glycosuria has been noted.

Duration:—The average duration of the cases collected by the British Chorea Committee was about ten weeks, but they may last from three weeks to six months. The rule is that the more severe the disease, the longer the duration. Recurrences are so common as to be a characteristic of the disease, females being more subject to recurrences than males. Endocarditis and pericarditis are more common complications of a recurrence than of the primary attack (11).

Treatment:—There are many types of treatment for Chorea now being used, some old, some new, but all embodying the cardinal principles of absolute rest and isolation. Children especially should be removed from the usual surroundings, preferably to the hospital and provided with absolute rest in a quiet, darkened room for about two weeks. Sedatives may be necessary during the worst stages and when this period is passed, tonsils and adenoids should be removed and also dental infection. Dwyer (4) advocates the following scheme in treating children:

1. Rest in bed in a quiet, darkened room for at least three weeks with no visitors, books or anything to hinder complete rest and relaxation.
2. For the control of symptoms during the acute stage, phenobarbital once or twice daily.

3. Warm tub bath when needed to promote relaxation and sleep.

4. Fowler's solution of arsenic after the most active symptoms disappear, for a period of two or three months.

5. Removal of tonsils, adenoids and dental infection as soon as the active symptoms subside.

6. The patient should not be kept in bed until all the symptoms clear up. Those with the residual symptoms can be taken out in a wheel chair in nice weather or allowed to walk about quietly when such effort is not tiring.

Arsenic has been used in the treatment of Chorea for over a century with results generally considered to have established its usefulness. However, authors differ regarding the dosage, some recommending large doses, and others small. Given by mouth, arsenic tends to cause vomiting and given subcutaneously, it causes considerable local irritation. Many authors have reported examples of Chorea treated by arsenic intravenously.

Graham (13) reports 45 cases treated with neo-kharsivan, liquor arsenicalis and sodium salicylate. His conclusions were:

1. That the course of Chorea would not appear to be influenced by treatment with arsenic. The improvement, if any, probably results from its tonic action and is not in proportion to the amount of arsenic given. Intravenous arsenic has no advantages.

2. Rest in bed and freedom from emotional disturbances will usually cause a disappearance of Chorea in
four or five weeks.

3. The administration of sodium salicylate is recommended in the hope that it might have some good effect on the rheumatic infection.

Among the numerous narcotics and soporifics recommended for the control of Chorea, luminol probably gives the most satisfactory results. Isoprol and chloretone are frequently administered as a soporific in England. These substances have a purely symptomatic effect; they are probably not able to prevent the development of endocarditis.

Nirvanol, another soporific, soon replaced these two preparations. It belongs to the barbituric acid group, and not only has their usual hypnotic action, but also seems to possess the power of producing a febrile reaction, and an exanthem more often than any other known drug of the kind when given over continued periods. Tisdall (14) describes 29 cases of Chorea treated with Nirvanol during 1929 at the East London Hospital for Children. Of these, 15 were considered good results, 5 were definitely failures and 9 were markedly improved. Six of the 15 cases classified as cures have relapsed within five months of the end of the series, while 9 or 31 percent of the total cases, remained free. The intensity and completeness of Nirvanol sickness varies in different cases. The curative effect in Chorea is apparently due to the bodily reaction produced by the sickness.

To produce nirvanol sickness, the drug is given orally in a dosage of 0.3 gm. per day to children of 3-14 years of age until the appearance of an exanthem. This interval of time is usually from 7 to 14 days after the administration of the drug is began. Rest in bed is ordered during the administration. Eosinophilia is a common blood finding in nirvanol sickness but the red cells and hemoglobin shows no alternation. Nirvanol certainly seems worthy of a trial, especially in the more severe cases of Chorea. It's effect on the ultimate prognosis remains to be seen.
Pilcher (15) recommends the use of phenyl-ethyl-hydantoin as a sedative. It has an action almost identical with that of nirvanol. Magnesium sulphate used intramuscularly is frequently used with good success. Daily injections of a 25 percent solution are given in 10 to 15 c.c. doses. Sometimes it is necessary to continue it 3 weeks or longer. If the disease becomes stationary, it is well to stop the magnesium sulphate and use typhoid vaccine. The vaccine is diluted with saline solution so that 1 c.c. contains 20 million bacteria. The dose is 1 to 2 c.c. and may be repeated in a week. There is a reaction for about 24 hours.

In 200 of Dwyer's (4) cases receiving magnesium sulphate injections only 2 were followed by swollen, painful indurations, and the majority were much benefited.

Vipond (16) and others advocate milk injections and it seems that this therapy has about the same effect as the typhoid vaccine, only being milder in it's reaction.

Rosenow has developed a serum which has been used in a series of patients without benefit. All the children so treated developed urticaria and other manifestations of serum rash.

Goodman (17) prepared a serum from the patient's own blood and reported excellent results. The patient's serum, about 10 c.c., obtained by centrifuging the blood, is given intraspinally after about 15 c.c. of cerebrospinal fluid has been removed. A reaction occurs similar to that occurring after injecting typhoid vaccine.

Charney (18) and Williams (19) each report two cases of Chorea treated with Small's antiserum. In two cases the equine concentrated serum was employed and in two the unconcentrated bovine form was used. None of the cases showed appreciable improvement after waiting a reasonable time. None of the cases had nor did they develop cardiac murmurs. This is the only significant fact which may, not with certainty
however, be attributed to the beneficent effect of the serum, but it is not impossible that four consecutive cases should be encountered without cardiac involvement. Small (9) himself wrote very convincingly so that many clinicians used his serum, but few have reported their results.

When a ketosis is produced, as is shown by the presence of acetone and diacetic acid in the urine, and becomes pronounced, it is found that choreiform twitchings in children usually diminish and gradually disappear. The ketogenic diet must fulfill the requirements of the child--caloric, protein, vitamin and salt---sufficient not only to maintain the body weight, but also to allow for growth. The diet usually accomplishes the desired results after several weeks. The problem of expense in the application of the ketogenic diet is of importance in selecting a method of treatment as fatty foods are comparitively high in price. Leopold and Rothstein (20) calculated their diet as follows:

Total Calories - body weight x 25 calories per lb.
Protein - 2/3 gm. of protein x body weight in lbs.
Carbohydrate is arbitrarily set at a figure, varying from 60 gms. to 100 gms., depending on the age and weight of the child.
Fat - Total calories minus the sum of the calories contributed by the P and C.

They employed this diet in a series of 12 cases of Chorea, with good results in 9.

Hiram Byrd (21) reports a case in which there was distinct evidence of pathologic currents registering in the motor nerves. The patient had experienced a drawing of the muscles of one side of the face and neck which was more or less constant. The arrest of this drawing immediately following the cocainization of the nasal ganglion and this was followed by the injection of alcohol. The patient has had no trace of Chorea for three years.
To judge critically a single therapeutic agent, one should eliminate other favorable influences which arise from hospitalization, adequate nursing complete rest and proper food; yet these measures are requisite for the patients welfare. The basic principles of quiet and rest, mental and physical, must always hold first place. It must also be borne in mind that Chorea is usually a self-limited disease which will recover without medication and that it is the chronic and most severe cases which come to the hospital.

Prognosis:- Ordinary cases usually result in a complete cure and but rarely a permanent loss of muscle power or changes in the mentality. The mortality in the cases of the British Chorea Committee was about 2 percent. The prognosis is more favorable in children than in adults and is better in a recurrence than in a first attack. The presence of severe mental symptoms or definite organic cardiac lesions justifies a guarded prognosis. If death occurs, it is usually due to myocardial failure or exhaustion due to lack of sufficient nourishment or lack of sleep or to injuries contracted during the characteristic violent movements of Chorea.

Pathology:- The pathological findings in acute Sydenham's Chorea have been so various that it may well be said there is no proved and definite lesion. Osler (5) stated in 1911 that endocarditis with involvement of the mitral valve is by far the most frequent lesion. Fatty degeneration of the myocardium with Aschoff bodies in the wall of the left ventricle and ulcerative endocarditis with metastatic emboli in other organs has been described.

Maclachlar (22), in a study of the capsular and pericapsular tissues of the tonsil in 18 cases of rheumatic fever and 5 of Chorea, found perivascular lesions consisting of lymphoid and plasma cells. In addition to this, atrophic muscle giant cells, fibrosis, bone and cartilage, and foreign-body giant cells in various parts of the tonsil were
encountered. An endothelial proliferative reaction of the lining cells of the capillaries and perivascular lymph spaces was the outstanding feature, in this study.

The outstanding findings in nerve cells have been chromatolysis, swelling and eccentric location of nuclei, complete destruction of some cells, deposits of fat droplets in the cytoplasm and in some of the perivascular spaces and a few small hemorrhages into the medulla near the restiform body. Zergler (23) thinks that many of these findings may be explained on the basis of post-mortem changes, but not the perivascular fat and medullary hemorrhages.

Lhermitte and Pagniey (24) report extensive pathological changes of the cerebral cortex and of the basal ganglia consisting of vascular infiltrations and cellular degenerations. They report a series of cases in which they found, at autopsy, marked vasodilatation and infiltration of the adventitial sheaths with mononuclear cells, lymphocytes, plasmaocytes and myelophages, both in the cortex and basal regions. They also discovered inflammatory reactions of the locus niger, intense lesions of the caudate, the thalamus, nucleus ruber, corpus Luysi and the perisylvian region. Of late, more significance has been attached to the lesions of the corpus Luysi. These authors agree with others of the Hughlings Jackson School that destruction of a region of the central nervous system cannot account for positive phenomena such as spasms, contractures, and Chorea-Athetonic movements; that all it can explain are phenomena of deficit. They accept, too, the idea of "diaschisis", in a general and dynamic way, whereby a lesion at one point may arouse inhibitions or releases therfrom, in centers at a distance. But when it comes to agreement with the idea, seemingly experimentally substantiated, that Chorea is due to direct pathologic change of the motor cortex, they object that the cessation of choreic movement in hemiplegia really proves nothing except that in hypertonic states such
movements are impossible. They do concur, however, in the idea that it is impossible to conceive of choreic movements without the existence of a cortex. This is shown by the absence of anything resembling choreic movements in decerebrate rigidity; also by the generally accepted view that such movements cease during sleep and hypnosis.

Conclusions:— According to the evidence presented in this article, it is evident that there is no uniformity of opinion as to the etiology, treatment or pathology of Chorea. Considering such irrefutable evidence which we have and the results obtained from various experiments discussed in this paper, I believe we are justified in adopting the following conclusions:

1. That there is much evidence that Chorea, rheumatic fever and endocarditis are very closely related.
2. That the mode of onset with varying febrile reactions, slow course with it's tendency to recurrences and heart and joint involvement and with mental symptoms resembling a toxic psychosis, suggests a bacterial or toxic origin.
3. That if such a specific organism or toxic product exists, it has not been conclusively demonstrated but in the light of Rosenow's and Small's work, it is probably a bacterium.
4. That the toxins formed from these organisms have an affinity for cardiac, nerve and joint tissue.
5. That rest, isolation and good nursing care are the most important factors in treatment, sedation being a secondary but necessary therapeutic measure.
6. That while Chorea is an encephalitis the exact location of the lesion or lesions in the motor pathways is not definitely known, but that the basal
ganglia, corpus striatum and its connection with the extrapramidal pathways and the Corpus of Luys, are undoubtedly involved.

HUNTINGTON'S CHOREA

Huntington's Chorea may be defined as a chronic, progressive, hereditary disease, appearing rarely before the third decade of life, and characterized by irregular choreic movements, speech defects and gradual dementia.

The disease was first described in 1872 by George Huntington of Pomeroy, Ohio, at the time practicing on Long Island. It was prevalent in parts of Pennsylvania and New York and was known to the laity by the name of "The Magrums".

Prevalence:- Cases have been reported from practically all European countries but Turkey. No cases have been reported from South America or the West Indies. Huntington's Chorea is extensively met with in the negro race, whereas, Sydenham's Chorea is rare.

Etiology:- The predisposing causes are sex, age and hereditary influence. Huntington found the disease more prevalent in males and the statistics of others bear out his claims. The disease occurs at any age, but usually in adults, beginning most frequently in middle age between the ages of 30 and 40. It appears that in certain families, the age of onset is earlier than in others and if the disease descends through several generation, it has a tendency to develop later in life.

The disease is transmitted from one generation to another but one generation may be passed over or its members suffer from some other form of nervous disorder as epilepsy or hysteria. If the children of choreic ancestors are so fortunate as to get through life with no manifestations of the disease, it is likely that the grandchildren and great-grandchildren will be free from the disease also. Grimbly and Wilson
report two cases of brothers who suffered and died from Huntington's Chorea at about the same time. The following points appear to be of interest in connection with the cases described:

1. The existence of a family history of insanity transmitted through the mother, three sons becoming insane and two of them developing Huntington's Chorea.

2. One brother had never been capable of regular employment but the other who was the younger had a long record of service in the British army prior to and during the World War.

3. The younger brother who had led an active life developed the disease at 34 while the elder who spent most of his life in an institution, exhibited no symptoms until he was 43.

Symptoms:— Abnormal mental status and twitchings may be noted several years before the typical coarser movements of Huntington's Chorea appear. Strong emotional disturbances and trauma seem to be factors in precipitating an attack. After the actual onset, there is an almost continuous play of involuntary movements giving rise to peculiar facial contortions, exaggerated gestures and peculiar speech. There are sudden tonic movements of the head, trunk and extremities which are somewhat similar to athetoid movements. The movements usually begin in the lower extremities and gradually work upward. The movements of the choreic limb may be voluntarily inhibited at the expense of greater activity of other muscle groups. As the disease progresses, the gait becomes ataxic, Phomberg's sign is often elicited and the deep tendon reflexes exaggerated, later becoming impossible to elicit because of the marked rigidity of the extremities. The superficial reflexes remain unchanged, the pupils respond to light and accommodation,
there are no sphincter changes and the blood and spinal fluid show no abnormalities. These are important points in the differential diagnosis.

The patients often early become absent-minded and later disorders of perception, memory and judgment appear. They frequently have delusions of persecution and often attempt to commit suicide. The mental changes are most marked toward the terminal stage of the disease.

The elder of Grimbley and Wilson's (25) patients exhibited the following condition upon admission to the hospital: Mentally he was confused in thought and could give little account of himself. He was restless, excited and resistive to attention. He had delusions of a persecutory nature regarding women stating that females followed him about and received money for ill-treating him. He had a marked inferiority complex. Physically he was fairly nourished, and the heart, lungs and alimentary system betrayed little abnormality. The knee-jerks were exaggerated, and he showed definite and constant choreic movements of the muscles of the limbs and face. The gait was very unsteady. The pupils were equal and normal in reaction to light and accommodation. Articulation was much impaired and he was defective in his habits. The patient showed a rapidly advancing mental deterioration and was always irritable and hostile in his attitude to his environment. His speech became altogether unintelligible and the constant and violent choreic movements of the head, neck and limbs resulted in many bruises and abrasions in spite of the use of a padded cell. He became gradually enfeebled and died three years after admittance.

Diagnosis:—There are five well-defined points which are usually found together but some of them may be absent.

1. Direct heredity.

2. Hyperkenesia characterized by sudden, purposeless, jerky movements of the head, face, trunk and extremities.
3. Onset in middle or later life.
4. Gradual progression with no remission of the symptoms.
5. Progressive mental deterioration.

In establishing a differential diagnosis, the various psychoses must be eliminated. Paresis is perhaps the most difficult to exclude because the two diseases often co-exist. Simple Chorea may be differentiated because of lack of history of a hereditary influence.

Treatment:— Prophylactic treatment of course would be the ideal method to control the disease. Some states have laws permitting sterilization of the mentally deficient but enforcement is very lax.

Other treatment is of little avail except to provide more comfort for the patient and relieve his friends from his care. The bromides, luminol and other sedatives combined with hydrotherapy are used to alleviate the choreiform movements.

Prognosis:— The disease has a slow and progressive course and a recovery has never been known. Death may occur as early as three years or as late as fifteen years after the onset. The patient may become bedridden and die from some intercurrent infection.

Pathology:— From a survey of the researches made by various investigators the pathologic basis of Huntington's Chorea seems to be a diffuse parenchymatous process involving the gray matter of the brain, particularly the caudate nucleus, giving rise at first to a degeneration and later to a disappearance of the small ganglion cells with but slight involvement of the large ganglion cells. In the mentally deficient cases, the cerebral cortex is found to have an involvement consisting of a pseudonuclear gliosis above the Betz cell zone, particularly in the frontal lobe and precentral gyrus.

Grimbly and Wilson's (25) cases, which we have previously discussed, presented the following pathology at autopsy: Both cases exhibited
thickening of the pia-arachnoid, local atrophy in the frontal and fronts-parietal areas, generalized cerebral and cerebellar softening, and a pronounced increase of cerebrospinal fluid with dilatation of the ventricles. The following micro-pathological changes were found.

1. In the cerebellum, pons and medulla, no changes except leptomeningitis.

2. In the mid-brain, caudate and lentiform nuclei, thalamus and precentral gyrus, infiltration of small round cells, most marked in the caudate nucleus.

3. In the thalamus, increased vascularity in addition.

4. In the lentiform nucleus and precentral gyrus, some degeneration of nerve cells.

Conclusions: -

1. Huntington's Chorea is quite rare.

2. Mental aberrations, choreic movements together with other cases occurring in the family are outstanding.

3. Differential diagnosis should be more carefully undertaken.

4. Treatment, except palliative, is of no avail.

5. The pathology consists of a degenerative process of the parenchyma with a characteristic localization of the lesions.

HEREDITARY, OR CHRONIC CONGENITAL CHOREA

Hereditary or chronic congenital Chorea is more apt to appear among those who are idiotic, imbecile or feeble-minded, but it occurs occasionally in those whose mentality is quite normal.
It may often, with good cause, be confused with Huntington's Chorea. Inasmuch as the treatment, prognosis and pathology are the same, no great crime would be committed in the name of science.

**SENILE, OR ARTERIOSCLEROTIC DEGENERATIVE CHOREA**

Senile or Arteriosclerotic Chorea occurs after middle life and is indistinguishable from Huntington's Chorea except that there is no family tendency toward the disease, and the movements and mental disturbances usually appear late in life. Mental deterioration occurs as part of the disease.

Treatment is the same as for Huntington's Chorea except that prophylactic treatment should begin earlier in life and tend to prevent arterio-sclerotic changes by a proper hygienic regime.

**CHOREA GRAVIDARUM**

Chorea Gravidarum, or the Chorea of pregnancy, occurs during or shortly after pregnancy.

**Etiology:** A pregnant woman who gives a past history of Chorea should receive immediate attention! Pregnancy favors a relapse in previously choreic individuals and the attacks are most common during the third month of the first pregnancy in women less than 25 years of age.

Whitmore (26) states that in England about 1901 the general opinion was that Chorea Gravidarum was a neurosis. This opinion was based by some vague inference on the appearance of Chorea Minor most frequently about the time of puberty, and of Chorea Gravidarum in young primiparae during their first pregnancy. It is easily seen that nervous shocks occurring in certain individuals at these two periods might precipitate a neurosis.

Of the other factors at play in the production of a neurosis and perhaps contributing to the development of Chorea Gravidarum, is the
heightened emotionalism at the time of marriage. Also the element of fatigue is not to be disregarded in pregnancy.

Symptoms: - The choreic movements exhibited are practically the same as those in patients with Sydenham's Chorea, but the movements may extend to the uterus.

Psychic manifestations are more marked in the Chorea of pregnancy, memory defects and maniacal outbursts being very marked. After delivery, the choreic movements gradually subside but the hallucinations and irritability may persist for some time.

Treatment: - The treatment is essentially the same as for Sydenham's Chorea, consisting of rest, isolation and sedation.

There has been considerable discussion as to the advisability of terminating pregnancy in severe cases. Kaffesieder's (27) and Stone's (28) results show that the patient does not suffer any by being carried through to term and the infant is given the advantage of the further development in the uterus. Hunnicutt (29) reports a severe case which failed to respond to the usual treatment and upon which he performed caesarean section. She spent a three weeks convalescent period at the hospital, gradually improving, but barely recalled her trip to the hospital and recalled nothing of her operation. The child was in good condition also.

Prognosis: - The prognosis varies directly with the severity of the disease. In severe cases, abortion often occurs, but in the milder cases, the child is usually carried to term. Of 438 cases recently reviewed, the mortality was 16.5 percent (11).

Conclusions: -

1. That an attack of Chorea minor usually precedes an attack of Chorea Gravidarum.
2. That there is a tendency to recur in succeeding pregnancies.
3. That it has a predilection for young primiparae in the first few months of pregnancy.

4. That emptying the uterus in Chorea of pregnancy need not always be the procedure of choice, but the patient should be closely watched for danger signs and intervention carried out if necessary.

**CHOREA ASSOCIATED WITH EPIDEMIC ENCEPHALITIS**

Chorea associated with epidemic encephalitis occurs as a part of the encephalitis. It is not so common as theoretically would seem probable. Tucker (2) recently reported a case in which the encephalitis began with influenza and was complicated by severe toxic hyperthyroidism and infected tonsils. Marked improvement took place after rest in bed, the administration of Lugol's solution, thyroidectomy and tonsillectomy.

Lesne and Langle (30) treated a child for Sydenham's Chorea which was a recurrence of an attack three years previously. The present condition began very insidiously; at no time did the patient present any lethargy or ocular disturbances, but there was a temporary enlargement of the parotids, and an increase in the sugar content of the cerebrospinal fluid. These two symptoms are important in epidemic encephalitis, and are not usually seen in Sydenham's Chorea.

It can be inferred from this case that Chorea can occasionally arise from epidemic encephalitis and be a symptomatic expression of it.

**CHOREA OF ACUTE INFECTIOUS DISEASES**

The Chorea of acute infectious diseases is also quite rare and is due to a complicating cerebritis. It may occur after or during the course of typhoid, diphtheria, scarlet fever, influenza, tuberculosis or any acute infectious disease.

Osler (5) states that scarlet fever with arthritic manifestations may be a direct antecedent cause of Chorea in 6 percent of all the
cases, and measles as a sole antecedent in 7 percent of all cases.

Cerebrospinal syphilis is a rare antecedent cause of Chorea but has been met with.

In concluding I will say that the ideas of the medical profession in regard to Chorea have undergone many changes of recent years. Originally Chorea was considered a functional condition with a family neurotic tendency in the background. It was then looked upon as a form of infectious disease of childhood, and later as an irritative phenomena of the cerebral cortex alone. During these periods, every conceivable movement, torsion and muscle phenomenon were classified as Chorea. These classifications have been largely discarded for the present view wherein Chorea is divided into seven forms, all of which are due to an irregularity distributed encephalitis involving not only the cortex, but also the basal ganglia and cerebellum.

In the matter of treatment, it would appear that rest and isolation, combined with various sedatives are still the most effective measures at our disposal, but that some of the newer synthetic drugs are worthy of more careful trial.

Prophylactic measures whereby the health of our young children is better conserved by periodic health examinations, proper diet and saner living conditions would no doubt reduce the incidence of Chorea tremendously.

The passing and enforcing of laws whereby the feeble-minded were, by some means, prevented from populating the earth with their kind, would, in one or two generations, abolish the chronic hereditary Choreas.

From a bacteriological and pathological standpoint, there is much to be done. Small, Rosenow and others have paved the way and it remains
for someone to definitely prove and demonstrate the existence of the specific organism of Chorea minor, if such an organism exists, and to prepare a specific curative serum.
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