

On Chorea

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Chorea is essentially a disease of the nervous system. The name “chorea” is given to the disease on account of the dancing propensities of those who are affected by it, and it is a very appropriate designation. The disease, as it is commonly seen, is by no means a dangerous or serious affection, however distressing it may be to the one suffering from it, or to his friends. Its most marked and characteristic feature is a clonic spasm affecting the voluntary muscles. There is no loss of sense or of volition attending these contractions, as there is in epilepsy; the will is there, but its power to perform is deficient, the desired movements are after a manner performed, but there seems to exist some hidden power, something that is playing tricks, as it were, upon the will, and in a measure thwarting and perverting its designs; and after the will has ceased to exert its power in any given direction, taking things into its own hands, and keeping the poor victim in a continual jigger as long as he remains awake, generally, though not always, granting a respite during sleep. The disease commonly begins by slight twitchings in the muscles of the face, which gradually increase in violence and variety. The eyelids are kept winking, the brows are corrugated, and then elevated, the nose is screwed first to the one side and then to the other, and the mouth is drawn in various directions, giving the patient the most ludicrous appearance imaginable. The upper extremities may be the first affected, or both simultaneously. All the voluntary muscles are liable to be affected, those of the face rarely being exempted.

If the patient attempt to protrude the tongue it is accomplished with a great deal of difficulty and uncertainty. The hands are kept rolling—first the palms upward, and then the backs. The shoulders are shrugged, and the feet and legs kept in perpetual motion; the toes are turned in, and then everted; one foot is thrown across the other, and then suddenly withdrawn, and, in short, every conceivable attitude and expression is assumed, and so varied and irregular are the motions gone through with, that a complete description of them would be impossible. Sometimes the muscles of the lower extremities are not affected, and I believe they never are alone involved. In cases of death from chorea, all the muscles of the body seem to have been affected, and the time required for recovery and degree of success in treatment seem to depend greatly upon the amount of muscular involvement. Romberg refers to two cases in which the muscles of respiration were affected.

The disease is generally confined to childhood, being most frequent between the ages of eight and fourteen years, and occurring oftener in girls than in boys. Dufosse and Ruzf refer to 429 cases; 130 occurring in boys and 299 in girls. Watson mentions a collection of 1,029 cases, of whom 733 were females, giving a proportion of nearly 5 to 2. Dr. Watson also remarks upon the disease being most frequent among children of dark complexion, while the two authorities just alluded to, Dufosse and Ruzf, give as their opinion that it is most frequent in children of light hair. In every case visiting the clinics at the College of Physicians and Surgeons of New York, and of which I have the notes, the subjects were of dark complexion. Temperature is said to exert an influence over the disease, it being according to some authors, most frequent during the winter months, and scarcely known in the tropics. Its mean duration is from thirty to sixty days, and although it is chiefly confined to children is not entirely so. Spontaneous terminations frequently occur upon the establishment of the menses in girls and of puberty in boys. There appears to be in certain cases of unusual severity or long continuance a degree of fatuity established, but it is usually recovered from upon the subsidence of the disease.

According to Rilliet and Barthez, as quoted by Dr. Condie in his work on diseases of children: "Patients laboring under chorea, being attacked by measles, scarlatina, variola, or other acute febrile disease of childhood, the chorea will generally be diminished in intensity or entirely removed."

They state that "out of nineteen cases nine were attacked with other diseases, and eight were evidently influenced by them." M. Ruzf, denies that concurrent diseases exert any influence upon the severity or duration of chorea. There is generally constipation with disordered stomach and appetite, it sometimes being defective and at others ravenous. Pain is not a common attendant upon the disease, although headache may frequently be present and sometimes tenderness along the course of the spinal cord, which is increased by pressure. "Epilepsy and hemiplegia," remarks Dr. Condie, "are not uncommon results of chorea," and in many cases falling under his notice the patients had died of tubercular meningitis. Dr. Todd states that "paralysis of a limb which has been affected by chorea is not uncommon." He also states that the sounds of the heart are often changed in chorea; a bellows sound is frequently observed, and is either aortic, systolic, dependent upon anemia, or much more frequently mitral, systolic or regurgitant.

Rheumatism and rheumatic pericarditis, have been shown by Dr. Copland to be frequent accompaniments to chorea. M. Lee, Begbei, Nairn, Kirkes, Trousseau, and others point out its frequent occurrence in connection with pericardial inflammation, and internal and external rheumatism. Trousseau says that examinations into the condition of the heart, and inquiry as regards rheumatism, should always be instituted.

As regards the pathology of chorea, very little satisfactory information has been gained, and indeed in a large number of persons who have died of chorea, and upon whom autopsies have been performed, in the hope of illuminating this dark subject, no morbid changes have been found of a nature to lead to the supposition that they were in any way connected with the disease, while lesions discovered in others, such as inflammation of portions of the brain, turgescence of its vessels, with effusion of serum; hypertrophy and injection of the brain and spinal cord; turgescence of the vessels of the brain and spinal cord, with several bony plates half way up the spine upon the piamater; a concretion in the left hemisphere of the brain; a tumor pressing upon the tubercular quadrigemina, with inflammation and sanguineous effusion; ecchymosis of the membranes, and a pulpy condition of the spinal cord, and an abscess within the cerebellum, as reported by Clutterbuck, Serris, Cox, Patterson, Roser, Willan, Copland, Monad, Hutten, Beight, Brown, Keir and Schrode, these might, I say, have exerted and doubtless did exert powerful influences over the cases in which they occurred.

The most probable theory, and one which I believe is most generally accepted at the present day is, that the disease depends upon some functional derangement in the cerebellum. Modern physiologists pretty generally agree upon the opinion first advanced by Flourens, that the function of the cerebellum is to direct, govern and coordinate the movements of the muscular system. This being the case, then, the irregular ungoverned movements of the muscles in chorea would most decidedly and emphatically point to the cerebellum as the seat of the difficulty. Undoubtedly, the ab[s]cess of the cerebellum discovered by Schrode was the exciting cause in that instance. But even if we take it for granted that we have discovered the sedes morbi we are still left in ignorance in regard to the nature of the derangement. And here we must leave the interesting subject of the pathology of this disease, and trust that the science, which has accomplished such wonders, through the never-tiring devotion of its votaries, may yet “overturn and overturn, and overturn it,” until it is laid open to the light of day.

The causes predisposing to chorea are various: Improper and indigestible articles of diet, confinement in illy ventilated apartments, with want of proper exercise; disordered digestion, etc. While the exciting causes are irritation from dentition, irritation in the stomach and alimentary canal; by worms, retained faeces, etc., anger, fright, rheumatism and injuries to the head. It is, also, singular as it appears, sometimes the result of mitation. Some authors mention the disease as occurring epidemically in schools, and in one instance among a religious sect in Kentucky and Tennessee. The disease sometimes assumes a character different from ordinary chorea, and a number of cases of this kind are alluded to by Watson. Some keep beating measured time as if they were marching to music, others are seized with an irresistible propensity to roll over and over; others to stand on their heads, others to walk forward or backward, sometimes rapidly and in one direction, until exhausted or checked by some obstacle.

The treatment of chorea now most generally adopted is by purgatives, tonics, counter-irritants, and anti-spasmodics. The first indication is, if possible, to remove the exciting cause and it will probably be different in each individual case. Bleeding used to be employed, and it is said with good results, but it is rarely used at present, except in cases when there is much pain in the head, or along the spine, when it may be taken moderately by cups or leeches.

Purgatives should be used to unload the intestines, and the bowels should be kept open, and in a condition as near normal as possible, not allowing a day to pass without a full and free evacuation. In the early stages the mild cathartics may be employed, as the fluid extract taraxacum and senna, calomel combined with rhubarb or jalap, and followed by castor oil.

The pil. comp. cath., or ol. ric., may be required when constipation is resisting, and the bowels are in a torpid state. Spts. turpentine is highly recommended by some, either by itself, or in combination with castor oil, tr. senna, etc.

Tartarized antimony has been recommended by some in as large doses as can be borne by the stomach.

In the *British and Foreign Medical and Surgical Review* for January, 1858, two cases are reported as being successfully treated by it. In the first case the chorea had lasted a month and was increasing in intensity. Large doses of tart. emetic were given on two successive days, and thirty hours after its first administration, all choreic movements had ceased. The chorea reappeared under a severe fit of passion, but again yielded to tartar emetic. In the second case the chorea was at first general, but immediately became partial. It resisted tonic and other treatment for six months, but yielded to tartar emetic in twenty-eight hours. Counter-irritation is often employed, either by blisters along the spine, pustulation by croton oil, or by friction with tartar emetic ointment, this last method being considered by some good authorities as preferable. The most essential part of the treatment consists in the administration of tonics, nearly all of which, both vegetable and mineral are found serviceable. Of the vegetable tonics, the best are the different preparations of bark, and the salts of quinia. Of the minerals, the chalybeates, arsenic and zinc, are undoubtedly the most valuable. Iron may be given in form of sesquioxide, proto carbonate or sulphate, and, indeed, will be found useful in any of its forms.

The proto-carbonate of iron, gr. v.–vii., given in syrup, will often be found very useful. The zinci sulph. has a great deal of testimony in its favor; the disease yielding to it when many other medicaments have failed. Its use is generally begun in small doses, say one grain, and gradually increasing a grain at a time until the stomach will bear twelve or fifteen grains. The effects produced in individual cases must guide the practitioner, and if one tonic fails, another must be substituted and persevered in as long as any benefit accrues from its use, and so on throughout the whole catalogue, if necessary. Cimicifuga, nux vomica and iodine have been used with marked success by some, and are highly recommended by them. Opium, belladonna,

hyoscyamus, stramonium, etc., are often serviceable in quieting muscular action and producing sleep, and the same may be said in favor of chloral hydrate and chloroform, the latter being used in event of failure of the other remedies. These drugs can be considered, however, only as adjuvants to the tonics. In conjunction with these means the cold bath or sea-bathing, plenty of exercise in the open air, and a strict attention to diet will in most cases prove successful.

Electricity applied along the spine has proven in the hands of some a powerful curative agent in chorea. It should never be applied directly to affected parts, as the disease is thereby aggravated, rather than relieved, and it should not be continued after the patient is convalescent. The diet should be nourishing and easy of digestion, the food to consist of such articles as beef extract, milk, eggs, etc., the condition of each patient indicating the kind and quantity of food to be given. Gymnastic exercise is often of much good and is employed by some as the sole means of cure. After recovery care should be taken to remove all exciting causes; the bowels are to be kept free and in a soluble condition, and in short, all irritation to both mind and body are to be removed as far as it is possible to do so.

And now I wish to draw your attention more particularly to a form of the disease which exists, so far as I know, almost exclusively on the east end of Long Island. It is peculiar in itself and seems to obey certain fixed laws. In the first place, let me remark that chorea, as it is commonly known to the profession, and a description of which I have already given, is of exceedingly rare occurrence there. I do not remember a single instance occurring in my father's practice, and I have often heard him say that it was a rare disease and seldom met with by him.

The hereditary chorea, as I shall call it, is confined to certain and fortunately a few families, and has been transmitted to them, an heirloom from generations away back in the dim past. It is spoken of by those in whose veins the seeds of the disease are known to exist, with a kind of horror, and not at all alluded to except through dire necessity, when it is mentioned as "that disorder." It is attended generally by all the symptoms of common chorea, only in an aggravated degree, hardly ever manifesting itself until adult or middle life, and then coming on gradually but surely, increasing by degrees, and often occupying years in its development, until the hapless sufferer is but a quivering wreck of his former self.

It is as common and is indeed, I believe, more common among men than women, while I am not aware that season or complexion has any influence in the matter. 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 again 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 78 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 belonged 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 50 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, at least 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 nowadays 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 incurables. Dr. Wood, in his work on the practice of medicine, mentions the case of a man, in the Pennsylvania Hospital, suffering from aggravated chorea, which resisted all treatment. He finally left the hospital uncured. I strongly suspect that this man belonged to one of the families in which hereditary chorea existed. I know nothing of its pathology. I have drawn your attention to this form of chorea gentlemen, not that I considered it of any great practical importance to you, but merely as a medical curiosity, and as such it may have some interest.