Medical.

CHOREA.

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THE name chorea, which for centuries has been applied to this disease, is most inappropriate. Its origin can be traced back to the Middle Ages, when occasional epidemic outbursts of combined mental and physical excitement occurred amongst the inhabitants of some parts of Germany. The victims of this hysterical dancing mania were ordered by the local magistrate to seek solace at the chapel of St. Vitus at Zabern. The saint's reputed power over the disease ended in his name being coupled with it, so that it became known as St. Vitus' dance. Two centuries later Sydenham's transference of the name, chorea, derived from the Greek *χορεία*—dancing, to an entirely different affection was a manifest error, but so closely has the association now become that the original application of the term has been comparatively obscured.

CAUSATION.

Rheumatism.—The frequent association of chorea with other rheumatic manifestations, especially carditis, has naturally led to this cerebral affection itself being regarded by most authorities as a direct sign of rheumatism. It does not necessarily follow that all cases of chorea are indisputably rheumatic in origin, and those who would doubt this have some evidence in their support. Nevertheless, for practical purposes of prevention and treatment, it is perhaps safer to regard every case of chorea as evidence of rheumatic infection.

Race.—Chorea is said to be rare in negroes and Indians, but then acute rheumatism as a whole is not very prevalent amongst these races. Both have been reported to occur from time to time in South Africa and India amongst the natives. In England the incidence of chorea among Jewish children is disproportionately high. The question arises whether their emotional temperament lays them particularly open to this nervous disorder, which is known to occur more readily in highly-strung, excitable children than in those of more phlegmatic character.

Age, Sex, and Social Standard.—Chorea is relatively uncommon in either sex below the age of 5 years, although the writer has recently seen a severe example in a boy of 2 years and 10 months. Above 15 years of age it begins to become less frequent and is rarely seen in the adult, except occasionally during pregnancy. The disease is certainly more common in girls than boys, in the proportion of about three to one, which is no doubt largely responsible for the higher incidence of rheumatic heart disease in the female.

The influence of heredity is not so evident as in other forms of rheumatism, but it frequently happens that more than one member of a family is affected in fairly close sequence. This has been urged as proof that chorea may be infectious, an unlikely contention when we consider how common it is in the wards of children's hospitals without spreading to other cases. Rheumatism is known to run in families and it is reasonable to suppose that certain members being of a similar temperament may fall victims to this particular form of it, if exposed to a common source.

As in other forms of rheumatism, chorea is much more common in the lower than in the upper scales of social life. Also it is encountered more frequently in urban than in country districts. The season appears not to have much influence, and here chorea differs from arthritis which is certainly adversely affected by sudden changes in the weather.

Upper Respiratory Infections.—The direct sequence of chorea to upper respiratory infections, notably tonsillitis, is not nearly so marked as in other forms of rheumatism. The same applies to exanthemata, such as scarlet fever, in which a throat infection is a leading feature. This no doubt accounts for the disappointing results obtained from tonsillectomy, an operation which appears to have little effect on the liability to subsequent attacks of chorea.

Debility.—Any weakening influence seems to favour the occurrence of chorea, and this no doubt accounts for its frequency in the lower classes. Anæmia has been especially cited as an important causative factor.

Pregnancy.—The relationship of pregnancy to chorea is interesting. It is more liable to occur in a young primipara during the first three months and, having thus once appeared, may return in later pregnancies. In most cases there is no other cause than the pregnancy to which it can be attributed, but a previous history of chorea or other form of rheumatism is not uncommon.

Emotion probably plays an important part. This is suggested by the onset of the chorea being most frequently seen when the patient discovers for the first time in her life that she is pregnant. An even more important observation in this connection is that of Sir Samuel Wilks, who found that a large proportion of the cases occurred in unmarried girls. Nevertheless, chorea of pregnancy is probably rheumatic in origin, although definite proof of this is not always easy to obtain.

PATHOLOGY.

From the available pathological evidence, chorea appears to be a meningo-encephalitis. The lesions, although widespread throughout the brain, are usually non-destructive in nature. Autopsies have shown that there is no extensive deposit of the infective agent. It would thus appear that the encephalitis is most likely due to a toxin, which in the first place might conceivably be derived from a microbic infection in another part of body. For the disease to arise, these toxins must fall upon suitable soil, which, from what has been said, would seem to be an unstable nervous system. This may be of congenital origin or induced by developmental changes or accidental causes, such as shock, great emotion, debilitating illnesses, or mental and physical overwork.

SYMPTOMS.

So well known are the symptoms that a detailed account is unnecessary. Analysis of the characteristic, spontaneous, purposeless movements has shown that atonia, ataxia and paresis are the principal factors involved, though they may not all be present to the same degree. At times the third of these is so predominant that a true paralysis is simulated. Poliomyelitis has even been diagnosed at times, a mistake which is more easily encouraged by the fact that in "paralytic chorea" one limb is exclusively or more markedly involved than others. This frequent predilection of the disease for one side of the body stops short at the limbs, choreic movements of the trunk and face muscles being bilateral although inco-ordinated. It is these sudden, ataxic, muscular contractions which give rise to the grotesque grimaces, the writhing movements of the body, the irregular movements or respiration, the clucking of the tongue, and, in severe cases, to the loss of speech.

A very characteristic action is for the hand quietly resting on the lap to be suddenly turned palm upwards. These are movements over which the patient has no control, but even with voluntary movements there is a lack of precision, and a relaxation of the contracting muscles, which is either premature or delayed. It is this disability which results in the proverbial dropping of the cup.

Some idea of the grade of the severity can be gained by inquiring into the patient's ability to hold things, to walk and to speak, since these functions become impaired in that order, the greater the cerebral involvement.

All the manifestations of chorea are aggravated by any mental excitement, but the movements cease during sleep, except in the very worst cases. Most cases show some mental change, which is manifested by emotional attacks and an easy irritability. Tears or laughter are invoked on the slightest provocation. More serious mental derangement has also been known to occur, amounting to melancholia, hallucinations, and even acute mania (*chorea insaniens*). It is of some importance to realize this before we attach any blame to a particular form of therapeutics. Fortunately, patients usually recover from such mental disturbances, however grave they may seem at the time.

The *temperature* in chorea is usually normal. Hyperpyrexia is a rare occurrence with a grave prognosis. The *pulse-rate* is not quickened unless the heart has become actively involved.

One of the most interesting recent findings is that in a large proportion both of acute and chronic cases the *sedimentation rate* of the blood is not raised. Here again is a strong suggestion that chorea is not always certain evidence of rheumatism, in which an active phase always produces a raised sedimentation rate. It is not inconceivable that those cases of chorea with a high sedimentation rate are the very ones in which the heart is liable to become involved. Could this be proved we should possess a valuable method of recognizing those cases which require special prolonged rest after their acute attack has subsided. It would also influence the prognosis.

DIAGNOSIS.

Most cases of chorea are so typical that no difficulty arises in making the diagnosis. With the present-day interest that has been aroused in all forms of rheumatism, there is a tendency to over-diagnose the condition, and in an endeavour to recognize it in its

earliest stages *tics* or *habit spasms* are frequently mistaken for chorea. This is all the more regrettable since the correct treatment for the one condition is likely to do harm in the other.

Great help may be obtained in doubtful cases by asking the patient to demonstrate the movements he is reported to be suffering from. Children with tics always have insight into their particular habit spasm and will delight in their voluntary demonstration. For the choreic child this is impossible, and on being asked he will be at a loss to repeat his movements and will often disclaim knowledge of them altogether. The type of muscular contractions is also different in the two conditions. In tics the same movement is repeated with an unfailing constancy and would seem in the first place to be the direct result of some irritation or trauma. In chorea, the movements are much more widespread, the contractions are irregular, dissociated, and on analysis could never have served a useful purpose.

Two other observations will serve to differentiate chorea. One is an inco-ordination between the diaphragm and intercostal muscles when the patient breathes deeply, and the other is a characteristic attitude of the hands when they are held out horizontally in front, palm downwards. In this position the thumbs will be pronated, the hands extended at the wrist-joint and flexed at the metacarpal-phalangeal joints, a contortion difficult to reproduce voluntarily. If then the patient's attention be distracted by asking him to protrude the tongue, typical choreic movements will probably make their appearance. None of these signs are met with in habit spasm and, unlike chorea, in this disorder the child's grip is firm and well sustained.

Mention has already been made of "paralytic chorea" and its occasional superficial resemblance to some true organic paralysis. A *monoplegia* coming on more suddenly than usual is liable to give the most trouble. Generally in chorea it is only a paresis, some movement can be elicited, and *poliomyelitis* can thus be excluded.

Hemiplegic paralytic chorea is again more strictly a paresis, never spastic and not involving the face. At the same time very slight spontaneous movements of other parts of the body are usually noticeable. Choreiform movements are only occasionally seen in *hysteria*, but here as a rule single muscles are involved, the onset is quite sudden and young adolescent girls are generally the victims.

The rare choreic type of *cerebral agenesis* (Little's disease) can be differentiated by its persistence from birth and some degree of associated spasticity. *Paramyoclonus* need hardly be mentioned since both sides are symmetrically involved, individual muscles are affected and the contractions are quite sharp and sudden, as if they had been stimulated by electric current.

COURSE AND COMPLICATIONS.

An average case of chorea can be expected to last for about six weeks, but it is not unusual for signs of the disease to remain present for two or even three months. Such persistence generally occurs in cases which have had a more gradual and less severe onset. They are more resistant to the usual forms of treatment.

A child who has once had chorea is always liable to relapse. This habit of the rheumatic infection to reappear in the same form is well known. Some patients may certainly have chorea during one attack and arthritis or pericarditis, for example, during another, but a repetition of the original type of rheumatism is more common.

Carditis or endocarditis may supervene during an attack of chorea ; this is strictly not a complication, but merely a spread of the disease to other areas of the body.

At times there is a synchronous multiple involvement, in which case the cerebral manifestations may be masked until the acute infection has subsided elsewhere. This is perhaps best seen in cases of acute pericarditis which have been preceded by chorea. Whilst the heart affection is at its height chorea is absent, and only becomes obvious as the pericarditis improves. The same applies to acute arthritis and chorea, which by a merciful providence of Nature rarely occur at the same time.

Findlay, in a long series of cases, found that when *carditis* supervenes, it generally arises during the first or second attacks of chorea. If these leave the heart unaffected, it is not so likely to become involved in any later recurrences. This effectively lays the bogey of the increased liability to cardiac involvement, the greater the number of choreic relapses.

Hyperpyrexia and mental complications have both been mentioned. It is the former which has provided most of the isolated cases that have come to autopsy in the past. Thrombosis of one of the big veins, such as the inferior vena cava, is a rare and grave complication.

TREATMENT.

Choreic children are highly excitable, and require calm and soothing nursing. They should lie flat in bed, and severe cases should be surrounded by screens in a quiet corner. It may be necessary to pad the sides of the cot, if the movements are violent enough to cause injury to the arms and hands. Sore places and whitlows are common on the fingers and may be very distressing, especially if they have been overlooked, and the child with loss of speech cannot draw attention to them. This aphasia is very upsetting to older children, and the nurse must try and reassure them and quickly interpret their wants. A warm pack is very soothing when the chorea is so incessant that sleep is disturbed. Various drugs, such as chloral, chloretone, antipyrin, and bromides are also useful sedatives. Chloral and chloretone must be used with care, as they are both powerful drugs, liable to poison the heart or the brain, if given in excess.

Cyanosis, tachycardia, shallow respirations, coma, and an erythematous rash are sure signs of an overdose. There is no risk of chloral or chloretone poisoning, if these drugs are only given in small quantities at first, and are omitted altogether if the child is asleep when the dose falls due. Proper evacuation of the bowels is also important. One grain of chloral hydrate for every year of the child's age, three or four times in the twenty-four hours, gives a rough estimate of the amount required. The dose should not be increased above 10 grains, even in older children, if it is to be frequently repeated. Chloretone should be given in somewhat smaller quantities, and an 8-grain single dose should be the maximum.

The insolubility of chloretone in water must be remembered, and if it is prescribed in solution, the bottle must be well shaken before pouring out the dose, otherwise toxic quantities will remain in the last few ounces.

Salicylate, or aspirin, certainly improves some cases, but unfortunately it does not prevent the rheumatic infection reaching and damaging the heart.

The amino derivative of antipyrin, pyramidon, is the most serviceable drug, and

the severest choreic movements soon diminish under its effect, without producing hypnosis, but the heart may still become involved.

Nirvanol (phenylethylhydantoin) has been widely used on the Continent and to a more limited extent in this country. The drug is given daily and stopped when a specific reaction appears, about the seventh to the tenth day, consisting of a morbilliform rash, pyrexia and eosinophilia; soon after this a dramatic improvement in the symptoms can be expected. In a few patients no sign of a reaction takes place, even after a course lasting fourteen days, in which case the treatment is of no avail, and should be discontinued.

Nirvanol treatment does seem to curtail the course of chorea and has been successful in certain long-standing subacute cases in which other measures have failed. Occasionally, however, even with a marked reaction it has failed to influence the chorea. The exact action of the drug is obscure, but there is some evidence to show that it whips up the patient's defences against rheumatic affections which are not being overcome. If that be so, one might expect failures in cases of chorea which may not be rheumatic in origin.

Nirvanol by some is regarded as a dangerous drug. This is only so if given in the wrong type of case, and then the danger lies not in any toxic property of the drug, if the correct dose is used, but in its liability to cause a temporary exacerbation of the rheumatic process before the Nirvanol reaction takes place and the desired improvement can be expected. This peculiar property might thus cause the flare-up of a smouldering pericarditis, or enhance the mental disorganization in certain severe cases of chorea. Nirvanol should therefore not be used in chorea in which mental symptoms are the predominant feature, or in cases complicated by acute carditis or occurring during the course of acute rheumatic fever. If these rules are observed the drug is perfectly safe and particularly valuable in overcoming cases of smouldering subacute rheumatism, either with chorea or active carditis, when other methods have failed and the patient is slowly losing ground. For further information on this form of therapy the reader can turn to the writings on the subject referred to below [1, 2, 3, 4]. Re-education of the muscles after an attack of chorea is sometimes useful and such occupations as knitting should be encouraged.

Provided the chorea has entirely disappeared the child may generally be allowed up eight to ten weeks after the onset of the disease, and at this stage convalescent treatment in the country is most advisable. Much longer periods in bed may be necessary if there is cardiac involvement, in the hope that permanent damage to the heart will be prevented. But even the utmost precautions are often powerless to stop the development of chronic cardiac lesions, which defy all known methods of prophylactic treatment.

REFERENCES.

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