A CLASSIFICATION OF CHOREA IN RELATION TO ITS CAUSES.*

BY

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OVER a period of many years a large number of cases of chorea have passed through my hands at the Royal Liverpool Hospital for Children at Heswall, and an opportunity has thus been afforded for investigating the causes of the disorder. Although chorea is classed as a disease and is described as such in the textbooks, it really is a condition of a neuro-muscular inco-ordination which may result from very different causes. It is a symptom rather than a disease, and I have come to classify it with reference to its origin, and not to mass all the cases under a single generic title. Thus we have:

1. The ordinary: rheumatic or toxic type.
2. "Stock-brained" cases: related to an inherited tendency.
3. A climacteric type.

There is often little to distinguish these cases so far as the movements of inco-ordination are concerned, but some of the associated phenomena present considerable distinctions.

One of the first lines of inquiry consisted in a primary classification of all the cases into those which were rheumatic or developed endocarditis or pericarditis and those which did not, and it appeared that the latter were often ascribed to fright or mental upset or some peripheral source of irritation, so that we had (1) rheumatic choreas, and (2) fright, etc., choreas.

Next came a careful scrutiny of the histories of both types, and the interesting fact was revealed that in the second class the children were sometimes left-handed themselves or, being right-handed, came of a left-handed stock, whereas the rheumatic children presented this kind of history in a very limited number of cases—probably representing rheumatism in children having the hereditary factor referred to. It was on this account that the non-rheumatic cases became styled "stock-brained choreas," and it is with these that I shall mainly deal here.

In a few instances children suffering from this form of chorea have belonged to families in which there were stammerers—a fact which throws some light on the nature of the condition. The choreic movements probably represent a gross motor ataxia or inco-ordination which may be regarded as analogous to vocal stammering. An example of this was seen in a boy (Robert S.), aged 6, who had well marked chorea, ascribed to the excitement of going to school for the first time. He was left-handed. The only other child, a boy, was right-handed and a stammerer. The father was a left-handed stammerer and so was his brother. Another similar case was seen in a girl who had chorea accompanied by acute vocal stammering which resulted from terror. She was left-handed and had a stock-brained history. I mention these cases simply to indicate the suggestive relationship which probably exists between this type of chorea and stammering, which has long been known to be in some way associated with familial left-handedness.

I have a large collection of cases of chorea presenting this stock-brained history, many of them acute, and so far as general symptoms are concerned in no way different from the rheumatic ones. The history is in some cases direct, in others collateral. It will suffice if I quote some examples of each to illustrate my point.

1. Margaret K., aged 9. Severe and prolonged chorea. Left-handed. Both parents right-handed. Four maternal uncles left-handed. This child had rheumatism two years before the attack.
2. John D., aged 13. Mild chorea; no history of rheumatism; attack followed a small operation. Left-handed. Has a right-handed sister. Father's family all right-handed. Mother, mother's paternal grandmother, father, and sister left-handed.
3. Robert S., aged 6. Severe chorea the result of fright; no rheumatism, no cardiac symptoms. Left-handed. Father left-handed and a stammerer, one brother right-handed and a stammerer. Father's brother stammered. Mother, seven sisters, and a second brother right-handed and normal.

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4. Henrietta H., aged 8. Severe chorea the result of being knocked down by motor. Right-handed. Father right-handed, mother left-handed. No history of rheumatism, no cardiac symptoms.

5. Edward J., aged 7. Severe chorea; no rheumatism or cardiac symptoms. Left-handed; stammered occasionally for three months before this attack. Mother's father left-handed.

6. Thomas E., aged 11. Severe chorea the result of accident. No rheumatism or cardiac symptoms. Right-handed. Father and mother right-handed. Mother has two left-handed brothers. Second attack of chorea followed dog bite.

7. Elsie C., aged 6. Severe chorea the result of dog bite. No rheumatism or cardiac symptoms. Right-handed. Father and brother left-handed.

8. Eileen A., aged 8. Severe chorea the result of fright (father threw paraffin on fire). Left-handed. Father left-handed, mother right-handed.

9. Hilda O., aged 8. Severe chorea—frightened by a dog. Right-handed. Parents right-handed. Has left-handed brother and aunt.

10. Muriel D., aged 6. Severe chorea—knocked down by bicycle. No rheumatism or cardiac symptoms. Right-handed. Mother's mother and brother left-handed.

11. Ada D., aged 12 (sister of above). Severe chorea—asccribed to sister's illness.

In all probability these choreas do not occur in children who come of a pure right-handed strain, nor would they in a purely left-handed one if such existed. Right-sidedness and left-sidedness are both hereditary, but the vastly larger proportion of right-handed people renders it impossible that there should be a pure left-handed strain in any family—that is, one uninfluenced by a right-handed dominance.

From the histories which I have collected I gather that it is where there is a partial or repressed transposition of cerebral function that we find the explanation of these cases—that is, they occur in children who are right-handed, but where there is a stock-brained left-handed influence at work, and in left-handed children where a repressed right-handed inheritance produces its influences. As a consequence of fright or other disturbance an unbalancing of co-ordination is brought about, owing to some unusual relationship of function on the opposite sides of the brain.

To revert to the stammerer, we have this kind of involvement of the speech apparatus. Stammering in a right-handed person may result from a determination towards left-handedness with a dominant left cerebral speech centre, whereas in the left-handed person it may result from there being a determination towards right-handedness with a dominant right cerebral speech centre. Hughlings Jackson, in his writings on the speech centre, regarded the centre in the left brain as being the leading centre, its fellow in the right brain as an automatic one.

It was on this basis that he accounted for the repetition of words and recurring utterances of sentences in a "barrel organ" way in some cases of right aphasic hemiplegia. He gave examples showing that these utterances often expressed the last call of the individual when struck down by the apoplexy. I recollect a case of the kind where the patient from the time of his seizure until his death repeated the words "I want some brandy"—no doubt the result of his feeling sick and faint when the haemorrhage occurred. Hughlings Jackson explained that these expressions probably come from the undamaged right automatic centre after being initiated in the left one immediately before it was put out of action. Thus there is a double cerebral service. If this be the case, it no doubt throws light on the relative functions of the two centres in a normal individual. One can well imagine that where a stock-brained influence impresses a tendency for automatism in a leading centre or for a centre having automatic functions to take on leading ones the inco-ordinations expressed as choreas might easily be brought about; the slender balance being upset under the influence of fright or other emotional or peripheral disturbances. Whatever the explanation, the fact remains that there is this type of chorea which occurs in children presenting a stock-brained history.

Before parting from this type I may mention three examples of congenital aphasia which have come to my notice. Two of them (girls) were left-handed, and all presented a collateral sinistral history. Of the two girls, one was mentally defective, the other, aged 6, was absolutely bright and quite up to the standard of intelligence

for her age. Although she could not speak, like the lesional aphasics, she could hum tunes and tried to sing with words. The third case was a boy (G. B.) aged 2 years and 10 months, who has never spoken. He makes efforts to speak like a child stammering very badly. He is a well proportioned child for his age, but his mental condition is difficult to estimate. His mother considers him quite intelligent. He is right-handed, but comes of a left-handed stock—that is to say, his mother is right-handed but her brother and sister and her mother also are left-handed. In these cases suggestion of lack of cerebral correlation again arises. I had one case of a left-handed boy who only began to stammer when an attempt was made to make him a right-handed writer.

A further outcome of this inquiry into the histories of the non-rheumatic choreas has indicated another causal factor which must not be overlooked. The condition occurs in both sexes, but the majority of the cases recorded were girls at or about the period of puberty presenting, as so frequently happens, certain constitutional disturbances manifested by the complexion becoming bluish or heliotrope, the hands and feet cold and blue, the heart large and palpitating, the liver somewhat small, and the thyroid enlarging. Here we have an unestablished balance of the internal secretions and a tendency to nervous upset. Chorea is not uncommonly related to this condition, and the cases constitute a class by themselves—the climacteric choreas. As in the stock-brained cases, the disorder is often precipitated by emotional causes. In the one it results from a chemical or metabolic inco-ordination, in the other from an anatomical one.

I am indebted to Dr. Simonne Thompson and to my resident physician, Miss Norah Walker, M.B., Ch.B., for having taken much trouble in tracing the stock-brained histories of every case of chorea, whether rheumatic or otherwise. Since leaving the Royal Liverpool Children's Hospital at Heswall, Miss Walker has carried out an independent investigation and has collected a considerable number of cases which confirm the above findings.

ACUTE APPENDICITIS IN A CHILD COMMENCING WITH DIARRHOEA AND CONVULSIONS.

BY

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THE case here recorded presents many points of interest. In the first place it had to be determined whether the child was suffering from meningitis with abdominal symptoms or from abdominal symptoms with meningismus, or possibly from two separate diseases. The one thing that seemed essential was to open the abdomen quickly and deal with the appendix. Presumably the very severe diarrhoea was an attempt on the part of the body to get rid of some of the toxin. The convulsive attacks also were apparently due to the same cause, for the operation seemed to produce an almost magical effect upon the appearance and condition of the child. Personally I have never seen an attack of appendicitis commencing with severe diarrhoea and convulsions, and I think that the case may be of interest to other clinicians; doubtless some surgeons have seen similar cases.

Maud A., aged 6, was admitted into one of my cots in the Royal Waterloo Hospital for operation on enlarged tonsils and adenoids. Four days after admission the child had a sudden very severe and continuous diarrhoea for fifteen minutes. The ward sister, who was with the child all the time, stated that she had never known a more severe one. For several days afterwards constipation followed. Shortly after the diarrhoea ceased general convulsions occurred; the whole body seemed to be affected, but especially the limbs and tongue. These attacks continued for four hours before they could be checked. There were twitchings of the lips, tongue, eyes, and limbs. The child at first appeared to be weaker on the right side of the body, but later the left side was noted to be weaker than the right. The pupils became dilated; the colour was first pallid, but later on the child became cyanotic. There was stated to be foaming at the mouth.

The urine examined during the attacks showed nothing abnormal. The child was given potassium bromide 10 grains and chloral hydrate $2\frac{1}{2}$ grains by the rectum soon after the convulsions began, but this had no result. Later 30 grains of the bromide and 5 grains of the chloral were injected, but ultimately chloroform

had to be administered to check the attacks. During the same night the attacks lessened and the child finally fell asleep, but later on was awakened by three or four attacks of vomiting. The vomit being of a greenish fluid nature.

The next day, when the child was examined, the tongue was very furred. The lips showed a small trauma from the application of the forceps to the tongue on the previous night. The child seemed to be in a semi-comatose condition. Vomiting continued. The bowels had not acted since the initial short attack of diarrhoea. Urine showed the presence of acetone, which possibly was due to the administration of chloroform during the attacks.

The child complained of pain in the upper part of the abdomen, and there was tenderness in that region. No tenderness was elicited by Mr. Cockell, my house-physician, at this period in either iliac fossa, but when he examined the child later in the day there seemed to be some tenderness in the right iliac fossa, which appeared to be a little more noticeable over McBurney's point than anywhere else in this area, compared with the left side. I saw the case a few hours later on the same day, and the child still appeared to be semi-comatose. The pupils were markedly dilated, the left perhaps more so than the right. Ophthalmoscopic examination was not made. Photophobia was present. No stiffness of the neck was elicited, nor discharge from the ears, nose, or throat. The tongue was dry and furred. The tonsils were distinctly enlarged, and very slightly red on the surface. There was slight stiffness of the left arm and left leg compared with the right side, and the left knee-jerk was also a little brisker than the right. On the left side there was slight ankle-clonus. Babinski's sign was not obtained. Kernig's sign was just present on the left side on this occasion only. The abdomen was retracted; tenderness was evident in the right iliac fossa in the region of McBurney's point. Resistance was noted. The note over the right iliac fossa was slightly impaired upon light percussion; there was no discharge from the vagina. Examination by the rectum disclosed nothing definite. The heart and lungs appeared to be normal.

The diagnosis of appendicitis was made, and Mr. Cairns Forsyth came down to the hospital and operated immediately. The appendix was found to be very long, distinctly distended, and acutely inflamed. It was filled with hardened faeces. A large number of shotty glands were seen in the mesentery. The child made an uninterrupted recovery. No further diarrhoea occurred, and the mental state cleared up wonderfully immediately the child came round after the operation. There were no more convulsions, and all the limbs appeared to be quite normal next day.

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PROCEEDINGS OF SECTIONS AT THE ANNUAL MEETING, BRADFORD, 1924.

SECTION OF SURGERY.

Sir CUTHBERT WALLACE, K.C.M.G., C.B., F.R.C.S.,
President.

DISCUSSION ON POST-OPERATIVE PULMONARY EMBOLISM.

OPENING PAPER

BY

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PULMONARY embolism may occur as the result of many different morbid conditions, but the most important, and that to which I shall chiefly confine my remarks, is as the after-result of an operation. Pulmonary embolism is by no means always fatal, and although no really reliable statistics on this point are available I should estimate that not more than one out of every ten cases of pulmonary embolism causes the death of the patient; in the remainder the patient recovers. There seems little doubt that most of the cases of so-called post-operative pneumonia are in reality cases of pulmonary embolism.

Pulmonary embolism may occur at any period after operation up to several weeks. The most common time is about the eighth or ninth day, but it has occurred within twenty-four hours of an operation, and in one case eight weeks later. Statistics are difficult to obtain, as the cases are comparatively infrequent and careful tables are not kept at most hospitals which will allow of the figures being