

CHRONIC HEREDITARY CHOREA.

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In 1842 Dr. Waters of Franklin, N. Y., wrote to Dunglison describing a peculiar form of chorea infesting certain families in that neighbourhood, which was remarkable in four points. It was markedly hereditary, it rarely appeared before adult life or after 45, it was incurable, and it in all cases induced dementia. Dunglison inserted the letter in his third edition, with the remark that he had never himself seen anything answering to this description.

The same disease was found by Dr. Charles Gorman of Luzerne, Penn.,¹ to prevail in other portions of the country also.

The next description is by Lyon,² who writes twenty years later that he has been familiar with it all his life, that it occurs in certain families only, that it is hereditary, and that it is called "migrims" by the people. He does not describe the disease in any detail, does not mention any mental affection as occurring with it, and in several instances states that it began in childhood. He gives three histories: (1) in which five generations, (2) and (3) in which three generations, were affected. Case No. 1 had two brothers who never had chorea, but one of them had two children "in whom well-defined chorea has existed for many years." He says, "To these cases many more might be added."

In 1872 Huntington³ described a chorea hereditary in a few families in a part of Long Island. It affected males more than females, came on gradually, always after middle life, was incurable, and was usually accompanied by insanity and a tendency to suicide. If one of the family escaped, his descendants were ever after free from it.

Ewald⁴ in 1884 described two cases in different families. There was the same strong hereditary character, but in neither case was the mind affected, nor does he mention insanity in any of the affected relations.

Clarence King⁵ gives a family history of disease in four genera-

¹ 'Inaug. Dissertat. at Faculty of Jefferson Med. Coll. of Philad.,' quoted by Dunglison.

² 'Amer. Med. T.,' N. Y., 1863, vii. 289.

³ 'Med. and Surg. Reporter, Philad., April 13, 1872, quoted by Sinkler in Pepper's System, vol. v. 440.

⁴ 'Zeitschrift f. klin. Med.' 1884, vii., Suppl. H. 51.

⁵ 'N. Y. Med. Journ.,' xli. 468, 1885.

tions and a description. He lays stress upon the insanity which accompanies it.

Peretti¹ gives an elaborate family tree of his case; the whole family springs from Frau N., who was choreic. Two children were healthy, and had healthy descendants; two were diseased. They had between them fifteen children, of whom nine had the disease and four had other nervous affections. Of the nine choreics, one had four children over 30, none of whom were affected, and three of these had healthy children themselves; four others had children, among whom one male aged 23 and probably two boys were already diseased. Insanity is only mentioned in four of the nine.

Huber² had a case of a man aged 38: the disease began at 30 in his face, spread first to the arms, then to the legs. He notices that the movements, which otherwise resemble juvenile chorea, are easily controlled. The only mental symptom in this case was weakness of memory. The man's sister, aged 42, was choreic and demented; his father choreic and insane, his father's sister choreic. The disease was found occurring in four generations.

In the Collective Investigation Report³ compiled by Dr. S. Mackenzie, West mentions a family in which three generations were diseased.

Zacher⁴ describes a case and gives the family history, showing three and perhaps four generations affected.

Hoffmann⁵ describes another set of relations similarly diseased.

Macleod⁶ treated and dissected two sisters, who were choreic and insane; their father had died of a disease which made him shake all over, and two brothers are said to be choreic. It is to be hoped that more details will be forthcoming; for though the cases seem probably to belong to this category, they are exceptional in more than one point, and the family history is insufficient.

Lannois⁷ traces the disease in four generations of one family, and describes six of the cases at length.

Ewald, Huber, Zacher, Hoffmann, and Lannois give most careful and excellent descriptions of the cases they observed.

From these authors a good picture of the disease can be constructed. It is a disorder of movement, affecting the voluntary muscles only, which exactly resembles chorea in appearance. Hoffmann does indeed describe a case (Jos. Kärcher) in which the movements were more like violent tetanic spasms; but it is so different in other respects also, that with all deference to Hoffmann I do not consider it right to class it with the present group. The disorder is hereditary, and affects more individuals of the families which it infests than most other hereditary diseases. Thus, taking only the patient's own generation, King has four affected

¹ 'Berl. klin. Woch.', 1885, No. 52.

² 'Virch. Arch.', cviii. 1887, 267.

³ 'Brit. Med. Journ.', 1887, i. 425.

⁴ 'Neurolog. Centralblatt,' 1888, 34.

⁵ 'Virch. Arch.', 1888, 513.

⁶ 'Journ. Mental Science,' xxvii. 1882, 191.

⁷ 'Rev. de Méd.,' vii. 1888, 645.

out of five, Peretti nine out of sixteen, Huber two out of two, Hoffmann five out of nine (Wipfler family). In the second generation of Lannois' family five out of twelve were choreic. These five had fourteen children, of whom eleven were diseased. On the other hand, all agree that when once the chain is broken the disease does not recur. An exception to this is, however, to be found in Lyon's paper; but he does not seem to be speaking from personal knowledge. The character of the inheritance is therefore peculiar. The disease affects both sexes, and is heritable either through the female as in Ewald's, or through the male as in Huber's case. It nearly always begins at or after 25 years of age, but occasionally earlier. Lyon, an exception in this as in many other points, speaks of its beginning at puberty. Peretti has a man of 23 in whom it had already begun to show itself, and another who had twitchings from childhood, and died paralysed at 19; while Felic. Wipfler (Hoffmann) began to be diseased at the end of her school-days. Twitchings in the face are noticed in children of diseased families by Peretti and Zacher. Waters places the age of onset not before adult life or after 45; Huntington says it never comes on until middle life; Ewald's cases nearly all began at 30; King places the onset between 25 and 35; Macleod's cases began at 60 and 70.

The disease does not develop out of ordinary chorea; but begins without known cause by twitching in the face (King, Huber), spreading thence to the arms and later to the legs, or is general (Hoffmann) from the first. The movement is violent, and coarse in character, not like Friedreich's paramyoclonus, a fibrillar twitching insufficient to move the limb. In the legs it produces a curious gait: there is a sudden stoppage, the patient looks as if he was going to fall forward, sways his body violently, at last is able to take a few rapid steps, and so recovers his balance (King, Hoffmann, Lannois).

It is stated in several cases that coarse motor power is preserved. In Felic. Wipfler it was but small. It is not uncommon for ancestors to be described as having been "paralysed." In some cases the finer movements are impossible, but it is noticed by several that these patients are much more able than ordinary choreics to control the involuntary movements by the will, so that those who are in constant and violent movement can yet drink, eat, and dress themselves without difficulty. In most cases the movements cease in sleep, but Hoffmann noticed them even then.

There is no wasting of muscles, no anæsthesia; the deep reflexes are natural or somewhat increased, and the electrical reactions are normal.

There is no heart disease connected with the affection, nor is rheumatism an antecedent. The bodily functions are carried on naturally, and micturition and defecation are, so long as the mind is sound, properly performed.

The disease is very commonly followed after a short time by some mental degeneration. This is noticed by every author except Lyon and Ewald. Though usual, it is not invariable. Two of Huber's cases were intelligent officials up to their death, and Ewald's cases were both perfectly sane. It not unfrequently

happens that other neuroses occur in members of the family who are not choreic (Peretti, Huber, Lannois). The choreic insanity begins with loss of memory and childishness, and goes on gradually to dementia. In two cases (Huber, Zacher) there were fits of violent destructiveness.

There have been three post-mortems recorded. Macleod (if his cases be admitted) found in one case a subdural hæmorrhagic cyst, with leptomeningitis over one side of the brain only, in another multiple tumours in the dura mater. Huber transcribes the post-mortem record of the father of his own patient. Pachy- and leptomeningitis were the only lesions. Another post-mortem, which will be of great importance, is to be expected from Klebe, who has dissected one of the cases which Huber reports in detail.

It is of the greatest interest to compare this disease with ordinary chorea. The movements are of similar character, and in both there is commonly mental affection. The one is closely connected with rheumatism and heart disease, and recovers. The other has no such connection and is progressive. The conclusion which the comparison suggests is that the two are produced by affections of the same parts, but of a different nature. On the other hand, it has certain analogies with general paralysis. Tremor and weakness in the one are replaced by loss of control and involuntary movement in the other; loss of muscular power supervenes at the end of some cases. The exalted delusions of the general paralytic are in some instances (Hoffmann) of this disease represented by cheerful misrepresentation of their real condition. The post-mortems show here, as in general paralysis, affections of the meninges.¹

That it is an affection of the brain is a generally expressed opinion, and that it is impossible as yet to say more is obvious.

¹ De Witt, 'Cincin. Lanc. and Observ.,' 1876, xix. 408, has a paper on bilateral chorea occurring in a general paralytic. I have not seen it, but it sounds as if it formed a link.