SOME FURTHER OBSERVATIONS ON FRIEDREICH'S DISEASE.

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It had been my intention in this paper to continue the digest of cases of Friedreich's disease which I published in a former number of 'BRAIN.'¹ But seeing that Dr. Judson Bury has recently, in his valuable article,² dealt exhaustively with the literature of the subject, I shall confine myself to recording some further cases which I have observed, and adding references to some papers which have appeared since Dr. Bury's publication.

My first set of cases are from a family resident near Hertford, comprising—Case I. A young man in whom the principal symptom has been progressive ataxia of the lower limbs. Case II. An elder sister, exhibiting the typical symptoms of advanced Friedreich's disease, plus sundry attacks of vomiting and of a syncopal (?) character. Case III. Their mother, bedridden with a spastic condition of the lower limbs.

CASE I.—John Brand, æt. 21 (at date of first observation), single, a carpenter by trade, became an out-patient under Dr. Bastian at Queen-square on Oct. 28, 1884. Dr. Bastian noted the ataxia, the absence of pains, and the comparative youth of the patient, and seeing the interest which I took in the case, very kindly transferred him to my charge.

He complains solely of weakness in the legs. He is a slight, somewhat pale and sallow man; gait unsteady, especially when he turns round; he is unsteady when he stands with his feet together and eyes shut. Hands said to be unaffected, but on closing the eyes, he cannot very readily touch the tip of the nose with his finger. Speech slightly hesitating and occasionally indistinct. Ocular movements normal; no nystagmus; pupils moderate size, act to light and accommodation. Optic discs normal. Legs fairly nourished, strength of muscles fair. Patellar tendon reaction absent on both sides. Cutaneous sensibility of soles and feet normal in all modes; no numbness or paræsthesia. Special senses apparently normal.

History of illness.—Was well till three years ago. Then had some sort of feverish attack, which he describes variously as "bilious fever," or "rheumatic fever." He was confined to bed for seven or nine weeks: was in a great heat and had pains all over him, but the joints were not red or swollen. On recovery he noticed the unsteady gait. He further seems to have had "a peculiar sensation" "which he cannot describe" from the legs up

¹ No. 25,

² 'BRAIN,' No. 35,

to the waist. The unsteadiness has gradually increased, especially during the last twelve months. A year after the illness mentioned above, he had scarlet fever, but this made no difference to his walking. Recently he has had attacks of nausea with retching, but he does not actually vomit, and the nausea passes off in three minutes or so.

Ever since the first illness he has been subject to pain in the limbs, which he thinks is rheumatic; it comes on only when he moves, and does not appear to have the characters of lightning pain.

Never diplopia; never difficulty in micturition (except occasional pain from "gravel"), nor of defecation. No history of syphilis. Ordered Syr. Ferri Iod. 3j; Pot. Iod. gr. v., ter die.

January 24, 1885.—Visiting him at his home, I was enabled to verify the observations made when I first saw him. There is now, however, no noticeable abnormality of speech, and he says himself this has improved. His hands he says are quite well, and he can do his carpentering work.

Farado-contractility of muscles in lower limbs tested and found normal. Plantar reflexes present. No deformity of feet. The teeth are peculiar—the upper incisors being small, somewhat set apart, the edges worn and somewhat honey-combed. The deformity is very slight, and suggests the so-called mercurial or rickety teeth, not those of congenital syphilis.

On *July 3, 1885*, he became an in-patient at Queen-square under Dr. Ramskill, and remained till the end of August. Dr. Ramskill kindly permits me to utilise the in-patient notes taken by our then resident officer, Dr. James Oliver.

The unsteadiness, which on admission as in-patient was worse than when I saw him in January, continued to increase. Slight unsteadiness of the head, and slight tremor of the legs was noticed. Nystagmus, lateral in direction, was observed, though only from time to time, and only when he looked to the extreme right or left. He complained sometimes of the legs feeling stiff, and of pains in the legs which seemed to draw them up, "cramping pains." There was slight hesitation and thickness of speech. He had some attacks of pain in the abdomen passing into his legs, and lasting a day or two; accompanied with nausea but no vomiting. A purgative given for one of these attacks upset him very much. Once he complained of a girdle-sensation. While in the hospital he was treated first with conium juice, and afterwards with quinine and iron.

July 30, 1886 (at his home).—His general health, which had rather fallen off while in London, has now improved. But the ataxia of the lower limbs has increased; he can now hardly get about the room unless he holds the furniture. He feels as if he had a sponge under his feet, or as if the floor were loose. Romberg's symptom is now well marked; thus when stripping for the examination of his chest, he has to sit down for fear of falling while he is pulling his shirt over his head. Beyond the ataxia, I can find nothing wrong with his lower limbs. The upper limbs

seem normal in every way; he can now touch the tip of his nose with the eyes shut. The speech, however, is rather indistinct, and the nystagmus more decided.

I have not seen him since, but a sister-in-law reports that his legs get no better, though in other respects he is well. He has now been taking nitrate of silver for a long period.

CASE II.—Linda Brand, sister of above patient, single, age 28 (1885).

January 24, 1885.—Seen at the family cottage; she was in bed on account of the severe cold. The history is partly from her own statements, partly from those of her mother, who was then alive.

According to her mother, she had weakness of the legs from the age of 5 years. It was noticed as she came home from school, that her legs became crossed, and that she staggered about. Eleven years ago (age then 17) she had scarlet fever, and this, to use her own expression, took her quite off her legs. She has been worse ever since. But she states that she can still stand and even walk a few steps with support. Ever since the fever too she has been unable to guide her hands, and her speech has seemed to her mother to be odd. With respect to this, she says herself that she has certain "attacks of vomiting which make her speech worse." In these attacks, she vomits phlegm for two or three days: anything that she takes would come up directly, and she therefore abstains from food. At these times also she aches all over, especially in the left arm down to the fingers. The attacks may come on, now, as often as once a fortnight, but less frequently in the summer than in the winter. They have been as frequent as this during the last three or four years, but she had them before that. During the last three or four years also she has had pains "like something biting her legs." During the last two or three years occasional incontinence of urine.

No numbness nor anaesthesia; never diplopia; sight said to be good, except for reading long together (error of refraction?). No deafness, at least to ordinary conversation.

Present condition.—She is of a pale sallow complexion, dark hair and eyes; mouth wide and slightly gaping, teeth prominent; the lower part of the face has a vacant expressionless look. (Her facial aspect strongly reminded me of another young woman with the same disease who was then under my observation.) Speech somewhat indistinct, words brought out with effort and pausing, syllables slurred sometimes. Tongue protruded straight, without tremor; slight tremor of facial muscles as she shows teeth. Well-marked nystagmus, chiefly as she fixes or follows; motions of eyes with this exception normal. Pupils moderate size, act normally to light and accommodation.

Upper limbs very thin, movements of hands and fingers (*e.g.* in trying to unbutton her dress) markedly ataxic; with eyes shut, cannot touch tip of nose with forefinger. No tendon-reaction on tapping radius.

Lower limbs thin; some muscular twitching in region of vasti interni just above knees, but chiefly, she says, "when she thinks

about it." The left leg she can scarcely raise from the bed; but she can raise the right, and tells me she can even bear weight on it when assisted to stand. No rigidity of legs. No deformity of feet. Tendon reactions absent at knees: plantar reflexes not obtained. Sensibility of feet (so far as I could ascertain) normal: muscular sense apparently normal—she knows how her limbs are placed in bed. Reaction of muscles to a moderately strong faradic current normal, but the current is felt very little in the left leg, albeit she feels the warmth and the touch of the electrodes. Spinal column (which could only be roughly examined) appears straight.

About eighteen months later, having heard that she was worse, and had become subject to alarming attacks of choking, I visited her again.

July 24, 1886.—She is now sitting in a chair, and is not in bed, but says she has grown so far worse that she cannot walk, and can only stand a very little. The expressionless look about the lower face is very marked; nystagmus unchanged; pupils as before normal; speech I think more defective, slow and rather drawing, words indistinct and yet somewhat separated; no definite tremor of tongue or lips. Further she says she has become subject to loss of voice: her voice she says "goes off." This is apparent as she speaks now, for sometimes as she begins a sentence, her voice gets squeaky, and subsides into a whisper before the end. There is now marked paralysis of the lower limbs; thus she can hardly move her feet off the stool (paralysis of flexors of hips and extensors of knees); neither can she dorsi-flex the feet or toes at all. I cannot however find any trace of deformity or contracture; she states that the toes "draw under," but this is probably the effect of the extensor paralysis. There is now I think actual wasting of the vasti, in addition to the leanness of the whole lower limbs. Sensation still quite normal. The hands are more ataxic than before; she cannot cut up her food, but can manage to feed herself with a fork. Complains of no pains. Has no trouble with the sphincters. So far as I can make out the spine is still straight, but the right scapula and the posterior part of the right ribs project in a manner that suggests a spinal curvature.

Coming to the "choking attacks," about which I had been told, she says that sometimes, and especially when she lies down in bed, she feels as if she could not breathe, and has to be held up. While talking to me something of the kind came on; she leant back in the chair with her head extended, then over one arm of the chair with her head drooping to that side, appeared to hold her breath, looked distressed and as if about to weep. Her pulse was easily felt at the beginning, but grew feeble towards the end of the attack. She came round in two or three minutes. No actual dyspnoea, no cyanosis, no loss of consciousness, nor any kind of convulsion. Except for the condition of the pulse and of the heart (see below), it might have been a sort of hysterical attack. These attacks she has had for six months, and worse since she was upset by her mother's death (in March of this year). The vomiting

attacks which she described to me on the occasion of my last visit still persist; the last visit was about three weeks ago.

An attempt to examine her larynx was altogether unsuccessful, as she was much too irritable and nervous.

On examination of her heart, I found well-marked evidence of disease, viz. apex beat just beyond nipple-line, in fifth interspace: action rapid: a systolic murmur, mostly musical, heard all over the præcordia, loudest at the left fourth interspace near sternum, and loud along all left edge of sternum, moderate at the apex beat and over aortic area, not audible behind. Probably this murmur was of very long standing, for ten years ago a medical man who examined her heart told her she "must have had rheumatic fever." She never had it to her knowledge. Other possible sources of cardiac mischief suggest themselves, either the scarlet fever, or what would fit in with the nature of her nervous disease—congenital malformation.

The father of these patients, William Brand, was at the time that I saw him (January, 1885) a healthy old man, who, with the exception of chronic rheumatism, had never had any illness in his life. His patellar tendon-reactions were normal. There appeared no reason to suspect him of alcoholism. About his ancestry I have no information. He died rather suddenly on June 8th, 1887, while sitting at table. He had previously, nevertheless, been suffering from dropsy.

More interest attaches to the case of the mother and to her family. I saw her but once; owing to her helpless state and the bitter coldness of the weather, it was impossible to make a thorough physical examination; but I was able to ascertain some facts of importance.

CASE III.—Mrs. Brand, age 64 (January, 1885). She gives a clear account of herself and her family. She was never ill till after her third confinement (age then 29 or 30), when she had a white leg. The present affection began to develop from twenty to thirty years ago (and therefore in no immediate relation to the white leg); she speaks of it before it laid her up as "a weakness." Twenty-five years ago she became subject to a feeling of loss of power in the right side; this feeling began in the great toe and passed up that side. It was momentary, but made her drop what she was holding; she has had it on and off ever since. For a period of eighteen years she attended at the Hertford Infirmary; she experienced difficulty in walking; she felt tired and used to fall about. She was also subject to "illnesses" which would take her quite off her feet. In these "illnesses" retching would come on suddenly, and last for some hours; she had abdominal pain at the same time. For some years, in addition to the weakness she felt a numbness right up to the navel; it felt as if she were walking on stumps. Sometimes she did not know when she passed a motion. This numbness has now quite disappeared. During all this period she had no notable pain.

Six years ago, an accident happened to her which seems to have formed a critical point in her disease. She stumbled and fell upon

her back, and has been much worse ever since. She has been unable to stand, and can only lie in bed or sit on a chair: she has also become what she calls "contracted"; the ribs of the left side are, she says, displaced, and she is subject to a dragging pain in the back and side. After the fall she used to have convulsions of pain in the right side, which seemed to "draw the side." From time to time she has difficulty in holding her urine; defecation normal, though the bowels are rather confined.

Present condition.—She is a healthy-looking woman in the face, though thin; at present is in bed. Speech quite natural. Sight and hearing said to be natural. No nystagmus: the state of the pupils unfortunately is not noted, but if I remember right they acted naturally. *Upper limbs.* Right grasp very feeble, left natural. No ataxia of movement; touches nose with finger-tip, even with eyes shut, fairly readily. Tendon-reactions present at both wrists. Right shoulder-joint stiff, cannot raise it above her head. She says this came on after the fall; whether it were due to adhesions, I did not determine. No rigidity of elbow, wrist, or fingers. *Trunk.* A marked projection of the ribs in the left inframammary region very suggestive of spinal curvature. Yet, so far as I can make out through her wraps, the vertebral column is straight. She says herself the back is not crooked, but the ribs she was told were displaced by the fall. She can sit up in bed when raised, but cannot get up into that position by herself. *Lower limbs* rigid, partially flexed at hips and knees, and the knees pressed together. She cannot move them from this position herself, and they offer resistance to passive movement. Tendon reactions present at both knees, and well-marked ankle-clonus present on both sides. The muscles both above and below the knees are small; I could obtain no faradic reaction, but had not a battery powerful enough to test this point thoroughly. Sensation roughly tested appeared normal in the feet. She has a dragging feeling in them which distresses her much, especially in cold weather.

This patient died in March, 1886; I suppose from some form of lung disease. She had had a bad cold and cough; and while convalescing from this sat out of doors in her chair. It was thought that she got a chill in this way, for she had to go to bed again, and died in five days' time.

I may say at once that I record the above case as being the mother of two typical cases of Friedreich's disease, herself suffering from some form of spinal disorder. I do not say that her symptoms were those of Friedreich's disease. There was no ataxia, no nystagmus, no speech-affection (although the disease was of long standing); and on the other hand, there was a spastic condition of the lower limbs, with preservation of tendon-reactions, and with ankle-clonus. It ought to be mentioned, however, that the woman thoroughly identified her own disease with that of her children; there are indeed some points in her history—e.g. the "illnesses" or vomiting attacks—which correspond with the history of her daughter's case, and it is by no means impossible that the fall on the back to which she attached so much importance may have

set up a myelitis calculated materially to modify her original symptoms.

But this much at any rate may be fairly argued, that the tendency to spinal disease in the Brand family came through this patient. For she was William Brand's second wife; and whereas by his first wife he has three living children all healthy, by this his second wife he has three living children, of whom two are paralysed. And again there is no record of nervous disease on his side of the family; but in her family was a marked tendency to paralysis. She gave me the following account of it.

Her father, William Savill, who died at the age of 70, had during the last twenty years of his life a "weakness" which she thinks resembled her own. He dragged one of his legs. His hands were unaffected. He used to complain much of "rheumatism."

Her mother had three "paralytic fits," in the last of which she died. These, so far as I could make out, were apoplectic: she distinguishes them, at any rate, from her father's illness, "which did not come on in this way at all."

She has two sisters of whom she has lost sight, but she never heard that they were in any way paralysed.

She had several (five ?) brothers. One of these died at middle age in a fit. Another, who died at 67, was for two years before his death "paralysed on the brain." He could not talk well, and his memory used to come and go. But he was never paralysed in his limbs, like herself.

I append a table of all the members of this family concerning whom I have been able to obtain information :—

Date, 1885.

The first two cases illustrate one special peculiarity of this disease, viz. its tendency to attack several members of one generation. The third case shows that their proclivity to spinal disease was inherited from the mother, having been in all probability inherited by her from her father. It need not perhaps surprise us to find symptoms of lateral sclerosis in the mother, as opposed to the symptoms of posterior sclerosis in the children, if we remember, that such knowledge as we possess of the morbid anatomy of the disease goes to show, that both lateral and posterior columns are affected in course of time, albeit the posterior are affected primarily and most completely. Other instances too are on record of spinal disease attacking several members of a family, yet under various forms.¹

Nevertheless it is an interesting question, whether this particular form of ataxy cannot, in addition to appearing as a "family" disease, i.e. among brothers and sisters and cousins, be shown also to be directly and strictly inherited. Now in a certain number of instances there has been reason to think that ancestors of the patients were ataxic;² but generally this has been a matter of history, not of the author's own observation, so that the exact nature of the ancestral disease remained doubtful. Vizioli,³ however, has settled the question in the affirmative. A man begat eight ataxic children. One of these cases Vizioli studied, and diagnosed as Friedreich's disease. This same patient of his begat two children, both of whom were under Vizioli's observation, and were the subjects of Friedreich's disease. Here therefore is evidence of direct transmission.

Taking "heredity," however, in the broad sense of family proclivity, there can be no doubt that it is a distinguishing feature of this disease—the feature indeed which chiefly led Friedreich to differentiate it from ordinary tabes. Nevertheless, as cases have been multiplied, it has become evident that the other characteristics of the disease are sufficiently definite to allow of a diagnosis even in sporadic cases; and further that, as might be expected, such sporadic cases do exist.⁴ It is important that this should be known, for it is in such cases (especially in their earlier stages, when perhaps difficulty of speech or nystagmus has not yet appeared), that mistakes in diagnosis are most likely to be made.

I have myself seen three instances of sporadic Friedreich's disease; the first under the care of Dr. Gee in St. Bartholomew's Hospital, the second under Dr. Hughlings-Jackson in the London Hospital, the third an out-patient of Dr. Howard Tooth's at the Metropolitan Free Hospital. This latter patient I have twice seen, once with Dr. Tooth two years ago, and again in the present year. Dr. Tooth kindly permits me to publish notes of the case.

¹ "Primary Spastic Paralysis and Pseudo-Hypertrophic Paralysis in Different Members of a Family." Philip, 'BRAIN,' No. 32.

² See cases by Carré, Rutimeyer, Brousse, Everett Smith, Judson Bury, and the author.

³ Vizioli, 'La Malattia di Friedreich,' 1885, pp. 11, foll.

⁴ See references in Dr. Bury's paper, 'BRAIN,' No. 34, p. 153; and also 'Archiv. de Neurol.,' March 1887, case by Blocq.

CASE IV.—Bridget Sullivan, age 19 (June 10th, 1885). A healthy-looking girl, though rather small in stature. Her chief complaint is unsteadiness on her legs. She certainly walks very unsteadily, looking at her feet as she goes. She cannot stand well with her feet together, and still less with her eyes shut. Even as she sits, there is a slight unsteadiness of her head. The hands are affected, but much less than the legs: she cannot do needlework as she used to do.

Lower limbs.—Absence of patellar tendon-reactions. Muscles well nourished: the quadriceps cruris contracts well, either when struck or under the faradic current. Sensation appears normal in every way. The left foot turns in a little, and there is here some tendency to club-foot. Otherwise no evidence of contracture.

In the *spinal column*, however, there is a lateral curvature, convexity to the left, in the dorsi-lumbar region; here also is some prominence of the spines (kyphosis?). Her mother too has noticed that she does not hold herself up when sitting. As regards the rest of the trunk, she has occasional pain, of a shooting character, in the left lumbar and iliac regions, but this does not appear to be a prominent symptom.

In the *upper limbs*, Dr. Tooth had noticed a certain amount of ataxia. Thus she could not, with her eyes shut, bring the tips of her forefingers together with certainty, nor touch the tip of her nose quickly.

Eyes.—Movements normal, no nystagmus; pupils act normally; fundus of both eyes natural. Sight is said to be impaired, but apparently she is hypermetropic. There is, I think, a slight peculiarity of *speech*, viz. extra distinctness of syllables and monotony of pitch; but at any rate it is too slight to have attracted the mother's attention. *Mentally* she was, when at school, intelligent and quick; but lately she has become irritable, and sometimes rude to her mother.

Menstruation has only recently commenced.

She has never had any fits, nor vomiting, nor headache, nor indeed any complaint but the present.

History of illness.—It began some two or three years ago. In May, 1883, she was in St. Bartholomew's Hospital, under the care of Dr. Gee. The following is an abstract of her symptoms.

"Ill for a year, with commencing tremors in the hands; drops things when spoken to crossly. Is getting worse.

"Pains in back. When a child, had abscess in the neck. Lateral curvature in dorsal region.

"Cardiac dulness above third rib. Apex-beat in fourth interspace within nipple line. Thrill (?) with apex-beat. Occasional harsh murmur with first sound over second left rib.

"No albuminuria."

A later note (May 2) says: "Tongue tremulous; right arm shakes. Diagnosis, chorea (?)."¹

¹ It seems to be a fact, that some cases of Friedreich's disease begin with symptoms resembling chorea; vide Medico-Chir. Society's 'Transactions,' vol. 68, pp. 149, 152; and Erlenmeyer, 'Centralblatt für Nervenkr.,' 1883, p. 385.

Since that period she has never been what she was before. The staggering gait was first noticed in November, 1884 (about seven months ago).

In June, 1887, I saw her again, after two years' interval. The chief change was a marked increase in the ataxia of the legs. She can now (it is noted) only walk when supported, or by holding on to the furniture. In doing so she sometimes gets her legs crossed, and there is certainly a tendency to throw the feet outwards and forwards. Her mother's expression is that she "throws her feet." She can stand with her feet apart, and can just stand with the feet together, but on shutting her eyes she would fall altogether. Sensation in the feet and legs is still perfect, and she knows their position when in bed. About the feet, especially the left, there is some bluish congestion looking like incipient chilblains. Left external malleolus prominent; left ankle-joint loose, and the foot tends to turn in. Left instep is high, and the toes decidedly tend to turn up. The right foot is well-shaped. I notice that her hands move awkwardly as she fastens up her dress, but she can do this, even fastening the strings behind her back. Sewing or fine work she cannot do, but she can "clean about" as well as ever. Tendon-reactions quite absent, whether at knees, wrists, or elbows. Speech not definitely affected, but it still seems to me a little odd. On fixing an object with her eyes, there is slight shaking of the head; and on fixing to the left, some (doubtful) nystagmus.

She is well-nourished, indeed rather fat; menses regular, breasts well-developed; heart and lung sounds normal.

The family history of this girl stands as follows (1887).

Mother, age 57, an active healthy Irishwoman; never been ill; never touches alcohol in any form, says a glass of beer would make her tipsy. Before marriage, was in good situations as domestic servant. Was one of seven brothers and sisters, none of whom were paralysed. Both her parents were healthy people.

Father (whom I did not see); also Irish, age about 58. Said to be "delicate-looking," but has never been ill. Is a confirmed drunkard. A pledge signed two months ago hangs on the cottage wall, but he drinks still. Comes home on Saturday nights "blind drunk, so that they cannot stop in the house with him." Yet it takes a great deal of liquor to intoxicate him. Sometimes, while in this state, he has pinned his paralysed daughter against the wall, frightening her very much. It should be mentioned, however, that in the two years preceding this girl's birth, he did not drink so much. His family history is unknown to his wife (who was my informant respecting him); but at least she never heard that any of his relatives were paralysed.

These Sullivans have had four children, viz.,

(1) Mary, died in 1867, at age of 2 years and 5 months, of "inflammation of the brain."

(2) Bridget, Case IV., given above.

(3) A son, age 19, healthy. Dr. Tooth examined him and found him free from ataxia, and his patellar tendon-reactions normal.

(4) Catherine, died in March, 1872, aged 1 year and 7 months, of "diseased mesenteric glands." She never walked.

Family predisposition is in this particular instance wanting. But as another possible factor in the ætiology, we ought to note the intemperance of the father. This point has been emphasized by other physicians, and I believe with reason. I have myself had the opportunity of studying Friedreich's disease in four different families, and in three of these, viz. Sullivan, just given, and Harmer and Woodcock, published in the 'Transactions of the Medico-Chirurgical Society,'¹ there has been a history of intemperance in the father. In the first two of these three there was no history of nervous disease among the ancestry; in the third (Woodcock) there was a strong nervous heredity on the mother's side; her father was probably ataxic, her sister insane. But in addition, her husband (as she has since informed me) was a hard drinker during their early married life, and her own notion (perhaps not an irrational one) is that her children's paralysis is due to the frights she sustained during pregnancy from her husband's drunken fits.

I find that parental or ancestral intemperance is mentioned in two instances by Friedreich, and also by Everett Smith, Vizioli, Quincke, Judson Bury, and Kingston Fowler. Some of the histories read almost as if intemperance had started a disease in the drunkard's offspring, which subsequently became hereditary. Thus the grandfather of Dr. Bury's patients had small feet, and could not walk in the dark, and his father again drank heavily; and in Vizioli's observations quoted above, we find that an intemperate man had eight ataxic children, and through one of these two ataxic grandchildren.

I admit, nevertheless, the force of the objections, firstly, that drunkenness may be itself the expression of nervous disease, and secondly, that the parental drunkenness may be a mere coincidence, for intemperance is very common, while Friedreich's disease is rare. But it is likely enough that several factors may be required for its production, and they may differ also in different cases. We know too little of the ætiology to neglect the study of any possible cause.

I have elsewhere remarked, that acute specific diseases appear sometimes to assist in the development of Friedreich's disease, or to cause an aggravation of it. The cases of John and Linda Brand (I. and II.) are curious in this respect. They both had scarlet fever about the same period of their lives, and after the first development of their nervous symptoms. John was none the worse for it; but Linda's disease was so much aggravated that she became a paralytic practically from that date. Nevertheless, John's disease developed originally after some febrile attack of uncertain nature. I should like to mention in this connection the case of two brothers recently shown to me by Dr. Hughlings-Jackson at the London Hospital. They both had Friedreich's disease; in the first the

¹ Vol. 68.

disease had developed in such close connection with diphtheria, that it was for a time questionable whether his symptoms were not due to some diphtheritic paralysis, of a permanent and progressive nature. Then the second brother came under observation, who had never had diphtheria, but exhibited nervous symptoms similar to those of his elder brother.

There is a condition not infrequently found in the subjects of Friedreich's disease which deserves notice—I mean the deformity of the feet. I do not think much attention was given to it before the observations of Rutimeyer, who noted the development of club-foot and a tendency to dorsiflexion of the toes. Dr. Bury describes and gives an illustration of the deformity. In the two Brands (Cases I. and II.) there was nothing of the kind; in the case of Bridget Sullivan (Case IV.) the left foot apparently is becoming deformed. I have watched the condition develop in another family (Woodcock). The instep becomes prominent, and the metatarsals appear to be shortened; at the metatarso-phalangeal joints the toes become over-extended, and at the first interphalangeal joints flexed. The foot looks humpy and shortened. Below, the plantar arch (at least, as seen from the inner side) is abnormally high, and the balls of the toes are very prominent. The foot also tends to assume the position of equino-varus. Perhaps a contracture of the long extensor of the toes associated with a paralysis of the interossei might produce the main features of the deformity. It certainly is not always present in Friedreich's disease, even in advanced cases (Case II.); and I have seen it in other diseases—once in a case of old-standing multiple neuritis, once in two sisters, who, though they could hardly be said to be suffering from Friedreich's disease, presented some symptoms which are worth mentioning in that connection. They were as follows—

A girl, æt. 19, was admitted by me to Queen-square Hospital for difficulty in walking. This, she told me, was due to a deformity of her feet. On examination I found the humpy shortened foot which I have endeavoured to describe above. *The patellar tendon reactions were entirely absent.* The difficulty in walking and the deformity of the feet had come on gradually about the age of 10, and she thought were getting worse. I could discover no other nervous symptoms. A sister, age 30, brought her, and this sister had a similar deformity, but in the right foot only. *The patellar tendon-reaction was quite absent on the right side, and only just present on the left.* The foot, she said, did not impede her walking unless she tried to go fast. On enquiring into their family history, they told me that a brother, aged 23, living in China, had the same shaped feet. There were six others in the family, reported healthy. Further, a maternal uncle had the same deformity of feet, but had no difficulty in walking.

I believe that cases of club-foot occurring in several members of the same family have been reported by orthopædic surgeons; perhaps an investigation of the nervous system might show, that in such instances there existed more than a mere local deformity, as I can scarcely doubt was the case in these patients.

FURTHER LIST OF PAPERS UPON FRIEDREICH'S DISEASE, OR
BEARING UPON IT.

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