Society Transactions

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A CASE OF SYPHILIS HEREDITARIA TARDA, WITH "TIP-TOE"

GAIT. DR. N. S. YAWGER.

The patient, who is from the service of Dr. Lloyd at the Philadelphia Hospital, is a girl, aged 10 years. The case is of some interest by reason of the tardy development of transmitted syphilis, but more particularly because it shows a very remarkable gait.

Present Trouble.—At 6 years this child began school and two weeks later she said her feet felt as though pins and needles were sticking in them; these pains kept up for a month and then she began to walk on her toes. Her behavior at this time was like that of other children—she talked, played, sang and recited pieces. At 8 years her speech became affected, she showed some disturbance of vision and was becoming noticeably deficient mentally. About six years ago, she developed epileptoid attacks; these occur once or twice in twenty-four hours and last from one to three minutes. In behavior she is irritable, bad-tempered and cannot be easily disciplined; if crossed, she gets into a tantrum—screams, kicks, bites and beats her head and body.

Previous History.—She was born at term and her infancy and early childhood was uneventful. She had a good start and apparently was a healthy child.

Family History.—The father is a laborer of about average intelligence and nonalcoholic. He admits a previous gonococcus infection but denies syphilis. The Wassermann test in the blood is negative. The mother died a few years ago—cause unknown. There had been but one pregnancy.

Examination.—Mentally, this girl is an imbecile; she is emotional, demonstrative and affectionate.

Station.—When standing her feet are rather widely apart and rest squarely on the floor; there is a backward bending of both knees, particularly the left.

Gait.—The child walks as though her extremities were rigid and usually, though not invariably, walks on her toes; she can keep up this gait for a considerable time and even run; the "tip-toe" gait is not due to talipes equinus; it is not constant, and she is not spastic.

Eyes.—There is no nystagmus nor ocular palsies. There is evidence of previous ulcers in the scars on the corneas. The pupils do not respond to light nor in accommodation.

Upper Extremities.—Judging from her volitional movements, they do not seem ataxic. The muscular power is good and the biceps and triceps jerks are equally and bilaterally exaggerated.

Lower Extremities.—These appear almost overdeveloped, both being unusually strong. At times, there is a bilateral patellar clonus; the patellar and

Achilles jerks are markedly exaggerated but there is no spasticity. There is an absence of ankle clonus and Babinski's sign. The sphincters are intact.

Sensory Changes.—Apparently there are none. She has typical Hutchinson's teeth and scaphoid scapulae. The Wassermann test in the blood was negative; two attempts at lumbar puncture gave no fluid.

Roentgenograms.—These were made from the lumbar spine and the knees; all were negative.

DISCUSSION

DR. J. HENDRIE LLOYD said that this child had been observed by him in the Philadelphia Hospital. Dr. Yawger had already called attention to all the features of the case. She has Hutchinson teeth very marked, and she has a rather characteristic history of the development of nervous trouble after she was born. She seems to have been a healthy child in early infancy and to have developed physical and mental deficiencies later on. That was in line with the history of hereditary syphilis as Hutchinson called attention to it many years ago. That is to say, the child may be born apparently perfectly well, and with the exception of some sniffles and some sores about the corners of the mouth and a transient keratitis, it may be practically a well child until 6, 7 or 8 years of age, when it begins to develop serious nervous disorders, such as epilepsy, paralysis and idiocy. This child has developed the ordinary appearance of the disease. The affection of her lower limbs, however, is peculiar. Her limbs are not spastic. She is fairly limber in her legs, but when she begins to run, up she goes on her toes as though by some spastic contraction of the muscles. She has quite typical Hutchinson teeth; also retarded mental development. The fact that the father has a negative Wassermann reaction and that the child has a negative Wassermann reaction has little or no significance. Dr. Lloyd believed in hereditary syphilis; it was commonly found that the child did not have a positive Wassermann in the blood or spinal fluid. This case raised the question as to what we mean by hereditary syphilis. According to the present-day biologists nothing can be hereditary which is not transmitted through the germ plasm. No acquired deficiency or deformity can be transmitted. That being so, and since we discovered that syphilis is a spirochetal disease, the question arises: Is there such a thing as hereditary syphilis? Biologists tell us that the spirochete is larger than the spermatozoon, it is larger than the ovum. If that is so, how can the spirochete get inside the spermatozoon? How can a spirochete be transmitted through the germ plasma? Hutchinson wrote of syphilis with remarkable prevision thirty years ago. He knew nothing about syphilis being due to a micro-organism, but he believed it was. He referred to the fact that some writers refuse to believe that hereditary syphilis can be transmitted from the father because of this very difficulty of seeing how the micro-organism could be transmitted through the spermatozoon. If that is true, hereditary syphilis is a misnomer. What we have in these children is acquired syphilis—acquired in embryo through the mother. In other words, it is congenital syphilis. Dr. Lloyd thought this child showed clinically fairly good evidence that she presents a case of retarded congenital syphilis.

DR. CHARLES W. BURR said that he had seen quite a few imbecile children who had very curious gaits that were not due to an organic disease, but were mental. They were habits that the child had acquired, and he rather doubted whether in Dr. Yawger's case the gait had an organic basis. Of course, the child was an imbecile and a syphilitic and all that, but Dr. Burr

was not convinced that it was due to a gross disease in the cerebral cortex. He would be inclined to believe, until the postmortem comes, that this was a trick in walking that the child had acquired. Imbeciles are very prone to all sorts of tricks, grimaces, dancing, etc. Dr. Burr thought it was quite possible that this condition instead of being a physical thing—he said he was speaking now of the gait rather than of other symptoms—that the gait may be mental rather than due to gross organic disease.

DR. AUGUSTUS A. ESHNER said he wondered whether one had not a right to ask for better evidence as to the syphilitic nature of this case? He thought that so-called Hutchinson teeth were not an infallible evidence. They were observed in association with other nutritional disorders, as has been shown. In the absence of the Wassermann reaction in the child and in the father, it seemed to be taking a liberty to describe this as a case of syphilis. The evidence was against syphilis rather than in favor of it.

DR. ALFRED GORDON said that the manner with which she stepped on her toes in order to walk reminded him of cases of extracapsular lesions. It resembled in one respect a case which he had named extrapyramidal hemiplegia and which he had exhibited before the Society. The moment the man attempted to move his hand, immediately there would be marked rigidity and spastic contraction of the extensor muscle. If he did not attempt to exercise the muscle the arm remained in exactly the same position as before. The moment an attempt was made to touch him or make active and passive movements, immediately the arm became rigid. When the child attempts to walk the flexor muscles of the legs become rigid. Here is a child mentally abnormal, which suggests some malformation of the central nervous system. The question in Dr. Gordon's mind was whether the extrapyramidal area was constitutionally abnormal so that stimulation of it produces rigidity of muscles controlled by that area. In regard to Dr. Lloyd's remark about hereditary syphilis, Dr. Gordon asked him to recall some facts reported in the literature in which some very young children supposed to have hereditary syphilis, had their spinal fluid and blood inoculated into animals, and spirochetes were found in the testicles. Dr. Gordon said he would then believe that even in the absence of the possibility of finding directly spirochetes in children they may be syphilitic through heredity.

DR. NATHAN P. STAUFFER said that if a nose and throat specialist might be permitted to ask a question, he had been wondering whether this child might have this trouble from infantile paralysis. The reason he asked was this: He had under his care some boys and at the time of the epidemic one of them died from the respiratory type of infantile paralysis and a number of boys who were in contact with that boy, and whom Dr. Stauffer was positive did not have disease of the vocal chords before, have since shown paralysis of the vocal chords. He wondered that in a number of the cases we see in which we are in doubt as to what causes the condition if the cases may not be due to infantile anterior poliomyelitis.

DR. G. LANE TANEYHILL remarked that apropos of a negative Wassermann in the blood serum in this patient (the mother not having been examined), this fall he saw a boy, aged 15 years, who is the physical size of a boy of about 11 years, and who had an undoubted case of *tabes dorsalis*. One of two undoubted juvenile cases of *tabes* Dr. Taneyhill has seen. The Wassermann was negative in the blood serum both before and after eight injections of arsphenamin, but the spinal fluid was positive—with 33-cells positive Wassermann reaction—and the colloidal gold reaction gave the paretic curve.

Both father and mother were examined; they revealed no signs of nervous disease and they cooperated far enough to have a Wassermann test made, but it was negative in both of them; but the spinal fluid of neither the father or mother had been examined. He thought it was very desirable to have the spinal fluid obtained from this child. That the blood Wassermann was negative did not exclude nervous system syphilis.

DR. N. S. YAWGER in closing the discussion, said in regard to the condition having resulted from infantile paralysis—unquestionably, the cerebral form could give rise to some of these symptoms; but here, the onset was slow and the child had many symptoms common to congenital syphilis including Hutchinson teeth, scaphoid scapulae and scars on the cornea. As to such teeth, these are typical of what Hutchinson described. It has already been remarked that a negative Wassermann is by no means uncommon in congenital cases. The comments by Dr. Burr on the gait, conform more nearly to what Dr. Yawger had in mind. Dr. Yawger had already remarked that the lower extremities were strong—appearing almost overdeveloped. Despite negative roentgenograms of the knees, there was an obvious defect at those joints, and he was inclined to believe that the gait was largely one assumed by the child to overcome the weakness at the knees.

A CASE OF MEDIAN NERVE INJURY, DUE TO STAB-WOUND
FROM ULNAR SIDE OF HAND. Presented by DR. T. E. SHEA
(by invitation).

Summary of Case.—The case presented is peculiar from the fact that a stab-wound in the hypothenar or ulnar side missed the ulnar nerve and injured the median. After a process of anatomic deductions, the injury was located in the median nerve at its emergence from under the anterior carpal ligament. Physiologically considered the deduction was made that the superficial fibers were injured, as it involved mainly the sensory distribution. Trophic disturbances were present which lead to thoughts, concerning the great factor displayed by normal nerve force in infections. The trophic disturbances in this case were compared to the limited gangrene of diabetes mellitus, and the belief expressed that ordinarily infection could not progress unless toxins destroyed the nerves controlling the protective forces of tissue.

DISCUSSION

DR. J. HENDRIE LLOYD said that trophic lesions following injuries to the median nerve had been reported. A case was reported some years ago by Haldor Sneve in which injury to the median nerve was followed by painless whitlows on the ends of the index and middle fingers resulting in loss of the distal phalanx in each of these fingers. Dr. Lloyd thought that was one of the risks in this case. Motion is not so much impaired in injuries to this nerve as might be expected.

DR. CHARLES S. POTTS said that he was considerably interested in the trophic disturbance in this boy's fingers because he had seen two cases of injury to the median nerve in which there was great liability to trophic ulceration in the skin area supplied by this nerve. In one of these cases a blister appeared on the end of the thumb. It finally dried up and left an area of sluggish ulceration which was very slow in healing and only did so as the other symptoms improved. The nerve was not divided, but injured.

Dr. Potts inquired if injuries to the median nerve were more likely to cause trophic disturbance of this sort than injuries to other nerves. Dr. Potts had also seen a woman the other day in whom an injury to the median nerve had caused symptoms similar to those of this man, and she had a little blister on one finger which looked as though it would break down and ulcerate.

DR. FRANCIS X. DERCUM said that one thing that would account for the freedom of escape of the ulnar nerve was the fact that it lies more superficially than the median. The knife entered well into the body of the hypothenar eminences and in that way the ulnar nerve lying practically above this level it escaped. The interference with the ulnar was very slight, if indeed any. Dr. Dercum asked the patient how much pain he noticed when it happened, and the patient replied, "No pain at all. My whole hand was numb."

DR. T. E. SHEA, in closing the discussion, said that in speaking of this case he had drawn the differentiation between the sensory and the motor fibers. Some authorities, which at the present time he could not quote, claimed that the superficial fibers of the nerve trunk were purely sensory fibers, while those which lie deeper were the motor fibers. If that particular fact held true the cut must necessarily have simply affected the superficial fibers of the nerve trunk. Speaking of the trophic disturbances, he mentioned the fact and asked what part the nerve force played and whether the trophic disturbance in this particular nerve involvement was not something like the diabetic gangrene we see.

HYSTERIA WITH MULTIPLE OPERATIONS. Presented by DR. FRANCIS X. DERCUM.

Previous History.—The patient was a woman, aged 27, from the wards of the Jefferson Hospital, and gave a history of five major operations for the relief of pain, none of which had been successful. The patient was unmarried; housemaid by occupation. No facts of importance were elicited in the family history. She had suffered from the ordinary diseases of childhood, had begun menstruating at the usual time, but at 17 had been nervous and had frequently suffered from convulsive seizures. At this time (1908) an appendectomy was performed. Subsequently (1910), a laparotomy for the removal of an ovary; in 1912, a gallbladder operation was performed. In 1915, an operation was performed for nephropexy, and in 1917 this operation was repeated.

Present Symptoms.—The patient presented no evidences whatever of any organic nervous disease nor of any visceral disease. While at the hospital, she had suffered repeatedly from hysterical convulsions and had complained of severe pain in the right side of the abdomen. At times, this was referred to the groin, to the right side of the abdomen. At times to the right lumbar region.

Examination.—This revealed marked areas of cutaneous painful hyperesthesia, but no tenderness or pain could be elicited on deep pressure. While Dr. Dercum could not, of course, claim that the operations performed had not been necessary, the patient herself stated that the symptoms from which she suffered had not been in any way relieved. It is more than probable that the pains of which she had complained and which had stimulated possible visceral disease had been purely hysterical in nature.

DISCUSSION

DR. CHARLES W. BURR said he thought it was a very common thing for hysterics to be operated on. Indeed, he sometimes thought that there were some surgeons who had no conscience at all. Also, in organic disease Dr.

Burr said he fancied that every man present had had at least one case of locomotor ataxia which had been operated on. At the Orthopedic Hospital about every eighteen months a patient comes to show a scar of an operation which was done in perfect good faith.

DR. SHERMAN F. GILPIN said that he had recently seen a patient who had been frightened in school at 14 and lost her voice. Since then she had had seven abdominal and two nose and throat operations in twelve years.

DR. ALFRED GORDON said this question of useless operations was very common. It was so ordinary that you look for it and as far as the conscience of the surgeon was concerned he would rather say that it was not their fault mostly. The patient is directed by the general practitioner to the surgeon with request for operation. The patient complains of pain in the abdomen. They find anteversion and all sorts of abnormal conditions of the uterus and refer all nervous trouble to that, to movable kidneys, to gastropexy and accordingly the patient is operated on; after the operation he is just the same as before. Dr. Gordon remembered one case several years ago which impressed him so much that he could never forget it: A young man of splendid appearance, who made the rounds of the hospitals for pain in the abdomen was treated in many ways. All sorts of diagnoses were made—dilatation of the stomach, and many others. He was massaged, even sounds were passed, cystoscopy was performed, and one surgeon advised the removal of part of the colon. Finally, he developed pain in the right side of the hypochondrium and was referred to a prominent surgeon who advised an immediate operation. Later in the evening the patient was put on the table; the patient being very emotional and excitable, saw the surroundings, physicians with white aprons and coats and instruments around. He became frightened, and jumped off the table and ran away. From that time he never had a pain.

DR. FRANCIS X. DERCUM said that he felt that his experience was not unique in this particular matter because there must be frequently recurring cases with multiple scars. Still, we must be very careful in regard to what we do and say. Dr. Dercum remembered many years ago when Dr. Osler sent him a patient who had an interesting case of abdominal pain. The man had pain in the epigastrium. He had been studied at the Johns Hopkins Hospital without any very definite result. He had acquired the morphia habit because of the pain, and when the man was sent to Dr. Dercum he withdrew all morphia, and then the patient became more than doubtful as to whether he had not a real organic basis for his pain. Dr. Keen operated and a small ulcer was found over the wall of the stomach. That man had an actual cause for his pain. That was not a case of hysteria. The diagnosis lay between tabes with gastric crises or gastric ulcer. Dr. Dercum finally persuaded him to have an exploratory operation performed. Dr. Dercum said he did not believe in exploratory operations as a rule. He thought this frequency of exploratory operations in nervous cases was a very serious matter.

TWO CASES OF FRIEDREICH'S ATAXIA APPEARING VERY EARLY IN LIFE. Presented by DR. FRANCIS X. DERCUM.

Dr. Dercum also exhibited two boys—brothers—aged 6 years and 4 years, respectively, both presenting the symptoms of a typical Friedreich's ataxia. The patients had been referred to Dr. Dercum's wards of the Jefferson Hospital by a nearby charitable institution.

Previous History.—The family history that could be elicited was rather scanty. However, it was learned that the father is living and is about 35 years of age; that he was unable to walk alone until about 9 years of age; that he comes from a strongly alcoholic family and is himself an alcoholic. Even when sober, he is said to walk as though intoxicated.

Present History.—The personal history of the older child is as follows: He has been apparently in good health aside from his present affliction. The patient had been abandoned by his parents in the early months of life and has been cared for in a public institution. He has only begun to walk during the past eighteen months. His gait is markedly ataxic; stands with his legs widely separated, and sways as though about to fall. The knee jerks are lost; there is a faint Babinski reflex on the left side, but no other feature of moment. The younger patient, aged 4, presents symptoms very much like those of his brother and equally pronounced; loss of knee-jerks, but no Babinski sign. Both of these cases are of interest because of the unusually early age at which the symptoms have made their appearance.

A CASE OF TABES DORSALIS SHOWING RESULTS OF TREATMENT, INCLUDING DRAINAGE OF CEREBROSPINAL FLUID.

Presented by DR. SHERMAN F. GILPIN.

This was a case of tabes dorsalis, showing effects of treatment by inunctions of mercury, with drainage of the cerebrospinal fluid.

History.—The patient, Mrs. A. K., a white woman, aged 49, was admitted to the Jefferson Hospital clinic in June, 1917. She is the mother of six living children and all of them in good health. Her present illness began four years ago with pain in left leg. Shooting pains in arms, neck and face have been marked.

Examination.—Physical examination shows typical Argyll Robertson pupils, diminished knee-jerks, and ataxia of gait and station. The ataxia was well marked and the patient so ill and weak it was necessary that some one accompany her on her visits to the clinic.

Laboratory tests on admission showed: Blood Wassermann, +; fluid Wassermann, ++++; lymphocytes, 48. Laboratory tests in March, 1918, showed: Fluid Wassermann, ++; lymphocytes, 43. During the last three months several Wassermann tests of the spinal fluid have been negative and no lymphocytes were found except at the last examination when the report is 4 cells per cubic millimeter.

Present Condition.—At present the patient feels well, has gained weight, stands well with the eyes closed and can walk the streets as well as she ever could. She has used mercury by inunction faithfully to the physiologic limit. Her spinal fluid has been drained thirty times.

DISCUSSION

DR. FRANCIS X. DERCUM said that it seemed at first sight difficult to account for the benefit of drainage in tabes and paresis. It occurred to Dr. Dercum that the explanation was as follows: If the pressure in the dural sac is increased, it must mean that less blood can flow into the cord than normally. If the pressure be suddenly reduced, a passive hyperemia of the cord probably ensues. You have here a method which is comparable to the Bier method in surgery. In the second place, if the spinal fluid is drained thoroughly, toxic

bodies must also be removed. Dr. Dercum said they took away 50 or 60 c.c. with great benefit. He was quite sure some of these patients would get much better even without the inunctions, just from drainage itself.

DR. GILPIN, in closing the discussion, said that Dr. Dercum had answered a question by Dr. Burr. He drained all the fluid that would run out. A week or two ago he took the fluid from a case of tabes for another physician and in a couple of days he reported to Dr. Gilpin that the patient was deaf. Dr. Gilpin saw her the next day. He could not make her understand by yelling in her ear. In three days she had recovered. Another thing—in taking fluid from a tabetic there is no following headache, whereas in the nonsyphilitic the patient has severe headache following spinal fluid withdrawal.

NYSTAGMUS ON MONOCULAR VISION. Presented by DR. ALFRED GORDON.

History and Examination.—A girl, aged 16, commenced to complain of poor vision six years ago. She improved under some treatment. Several months ago there was a return of the condition. When she came under the author's observation, the following symptoms were noticed: On monocular vision nothing abnormal was present. When one eye was covered the other eye immediately assumed the state of lateral nystagmus with the quick movement externally. Moreover, when the patient having both eyes uncovered would turn them externally to the extreme angle, either eye could not remain fixed in the external angle but would tend to move away. The pupils and eye-grounds were all normal. The patient also presented some atasia of the left hand in the pointing test and diminished knee-jerk on the left side.

Treatment.—Although the Wassermann reaction of the blood was negative, nevertheless, she was placed on mercury and iodids. There was considerable improvement in the power of the external recti muscles to hold on when the eyes were placed in the extreme external position, the atasia of the hand disappeared totally. The nystagmus on monocular vision remained unaltered.

Comment.—The condition of the eye-globes—namely, the nystagmus and the inability of the external recti to remain in a fixed position—point either to a congenital weakness of the latter muscles or to a lesion in the muscles of both sixth nerves.

DISCUSSION

DR. FRANCIS X. DERCUM said that the little girl looked to him as though she had rather prominent eyes, rather a large thyroid, and rather a rapid pulse. Have we not, perhaps, here a case of hyperthyroidism or exophthalmic goiter associated with the nystagmic involvement?

DR. WILLIAM G. SPILLER said that in examining the patient he had observed a phenomenon which Dr. Gordon had not mentioned. Dr. Gordon had stated that the external recti muscles were weak, but Dr. Spiller had noticed that this weakness like the nystagmus was distinct only in monocular vision. When the girl looked to the right or left with both eyes opened the weakness of the external recti muscles was not evident, but if she closed one eye the outward movement of the other eye was very imperfect and the eyeball could not be held well in the outer canthus. It was evident that the innervation of each external rectus muscle in binocular vision was greater than in monocular vision, and probably the innervation of the internal rectus muscle of one eye

caused an overflow of innervation to the external rectus muscle of the opposite eye. While this double innervation might occur to some extent with one eye closed, it was greater with both eyes open, and was then sufficient to overcome the slight weakness of the external rectus muscle, which possibly was congenital.

The nystagmus occurred under precisely the same conditions, and it was reasonable to associate one phenomenon with the other. The nystagmus occurred only in monocular vision probably because of the weakness of the external rectus, which being slight did not prevent the eyeball from moving outward, but was sufficient to prevent it from being held firmly; and in the attempt to fix the eyeball repeated innervation was necessary and a pseudo-nystagmus was thus produced. A pseudo-nystagmus may readily be observed in a person looking out of the window of a rapidly moving train, as the eyes are rapidly losing from vision the object fixed and a new object is equally rapidly coming into the visual field.

DR. G. LANE TANEYHILL said that he would suggest that the weakness of the external rectus was not sufficient to give the patient a squint. It was weak only enough to cause a little swinging inward on monocular vision which she tried immediately to correct, producing "nystagmus." This suggestion accepts Dr. Spiller's observation.

DR. DERCUM said that nystagmus was a positive sign. It was not a sign of mere muscle weakness. A muscle may, at times, present a nystagmoid movement which comes to rest after a while, but is a positive symptom.