LOCOMOTOR ATAXY OCCURRING IN A YOUNG WOMAN

TABETIC ARTHROPATHY WITH DISLOCATION OF BOTH HIPS;
SIX PREGNANCIES OCCURRING IN THE COURSE OF
THE DISEASE

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LOCOMOTOR ATAXY accompanied by arthropathy of the hip-joints and occurring in a child-bearing woman is sufficiently rare to warrant the detailed record of a case. The following patient has been under observation for two years, in which time two pregnancies have been passed through. Previous to the time of coming under my notice the woman was under the care of Dr. Wade and of Mr. Barling at the Birmingham General Hospital. During the last two years the affection of the nervous system, although there have been vicissitudes, has remained nearly stationary, so far as can be determined clinically.

Mrs. A—, æt. 29, came to the General Hospital, Birmingham, on March 13th, 1894, complaining of pains about the lower abdomen, the sacral region, and across

the hips, and that her urine had been dribbling away since the beginning of January. The last menstrual period had ceased on January 2nd. On examination there was found remarkable deformity about the hips and loins, as detailed below. Abdominal examination showed the bladder to be distended, the organ reaching the umbilicus and forming an oval, moderately tense tumour, with long axis directed upwards and to the left. *Per vaginam* the cervix uteri was found low down and in the axis of the vagina; the body of the uterus, also in the axis of the vagina, was enlarged, rounded, and softened, and the long axis of the uterus ran to the right as well as backwards towards the hollow of the sacrum. The plane of the pelvic brim looked almost directly forwards. A little more than a pint of slightly turbid urine was withdrawn by catheter, and the uterus was then easily returned to its natural position. The diagonal conjugate diameter measured four and a quarter inches.

On March 16th the patient reported herself as feeling very much better. There had been no trouble with micturition since the previous visit. The uterus was now entirely above the pelvic brim, and extended as high as the umbilicus; it was as large as is usual at the third month of pregnancy, and was displaced bodily to the right, its long axis lying altogether on the right of the median line, and running somewhat obliquely from above downwards and towards the middle line. On vaginal examination the os uteri was found to be high up, and the cervix partly softened. There had been no vaginal discharge.

On this date, March 16th, a careful examination was made, and notes taken on which the following history is based. The patient had previously been in the General Hospital on two occasions,—in October, 1890, under Dr. Wade, and in July, 1891, under Mr. Barling; advantage was taken of the notes made on these occasions to control the woman's statements, and these were found to be remarkably accurate in almost every

detail. Until the age of sixteen, the patient was always very strong, and there appears to have been no infantile disease of importance. About sixteen there began to be shooting pains in various parts of the body, but especially in the lower limbs, and since then the patient has not been able to get upstairs without resting, because of pains in the bones; when the pains were bad the veins swelled. The catamenia began at eighteen, and since then have been regular monthly, lasting about five days, moderate in quantity; there has been no special pain, but the general shooting pains have been worse at the periods.

Mrs. A— was married December 26th, 1887; at that time she was straight-backed, and her height then and for a year afterwards was 5 feet $1\frac{1}{2}$ inches. The present height, ascertained by measurement, is 4 feet $7\frac{1}{2}$ inches, so that the patient has lost six inches since 1889.

The first child was born in October, 1888, and was suckled for two months; during this pregnancy the woman says she was well except for "falling of the womb." The labour was natural.

The second child was born in November, 1889, and suckled for two months; during this pregnancy there was "falling of the womb" all through, and much neuralgic pain about the abdomen. On getting up the patient was bent forward, and could not straighten her back. About a month after the confinement the left knee became swollen, and remained so until the third baby was born. The onset of the swelling was accompanied by much pain in the joint, which, however, soon disappeared. About the same time there was creaking on movement of both hips and both knees; the general strength failed, but she did not become shorter until the third pregnancy.

In October, 1890, she was in the General Hospital under the care of Dr. Wade, when it was noted that the shooting pains had been much worse in the preceding twelve months; at this time the pains, which had

formerly been seldom and of short duration, lasted with short intermissions for hours and even days; they were worse in the legs, and would cause the patient to fall down if standing; when severe, they were followed in about an hour by a pain in the cardiac region, causing a fluttering of the heart. About three months before this, that is in July, 1890, patient had the "dropsy;" the water gathered in about six days, and the doctor is said to have removed six pails of water from her by tapping; the patient did not keep her bed, and there has been no return of the dropsy. While under Dr. Wade it was noted that the woman was five months pregnant. The chief points at this time in the condition of the nervous system were slight anæsthesia in both legs, loss of knee-jerks, slight nystagmus, complaint of dulness of vision, difficulty in standing and walking, and severe pains. There had been tired, aching pain, and loss of use of the right leg for a fortnight; the same leg always felt cold and numb above the knee; it swelled at night; even when thoroughly warmed at the fire, the former numbness quickly returned. There was swelling about the right great trochanter, and when the patient stood on the right leg there was marked lordosis. A lateral curve of the upper dorsal spine was also noted at this time. The pupils were observed to react both to light and accommodation.

This third pregnancy terminated in natural delivery in March, 1891; a month before the confinement the patient had a severe fit; there was a second severe one at the end of 1891, and since then there have been many slight ones in which speech and sight, and sometimes consciousness, have been lost; the mouth worked, but the mother of the patient used to prevent tongue-biting with a spoon placed between the teeth; urine has never been passed in an attack.

In July, 1891, the woman was admitted under the care of Mr. Barling with tabetic arthropathy of the right hip-joint; the limb had gradually shortened, and was now

two and a quarter inches shorter than the left one ; the swelling of the left knee was much less, but there was still some thickening around the margin of the tibia and femur. There was a small, freely moveable piece of bone connected with the right anterior superior iliac spine. Sight had improved, there was still slight nystagmus. The lameness had increased, and the gait was peculiar and characteristic.

Shortly after leaving the hospital in September, 1891, the woman says a raised boot was made for the right foot, the leg having become four inches shorter than the left : but before she could wear it the left leg suddenly, without any pain, became as short as the right.

In 1892, there was difficulty in telling the position of the feet, the patient not being able to tell, for instance, whether they were in bed or out of it, without looking at them. In the same year she was cross-eyed for about three months.

The fourth child was born in September, 1893, and was suckled for about a month ; during this pregnancy the patient did not feel at all worse. There have been two monthly periods since this labour, the last ending on January 2nd last. For nine months there has been occasional difficulty in holding and in passing urine. The bowels have varied, sometimes being very stubborn, and sometimes acting regularly ; in the last six months they have on a few occasions moved without the patient's knowledge.

The woman has observed no difference in the character of the pains in her various confinements, which have all been natural and easy ; the third and fourth were, if anything, easier than the first two ; the fourth was over before the doctor could reach the house ; there have never been any after-pains.

The second child died when ten months old of "diarrhoea and dysentery" ; the third at two years of "inflammation of the lungs, bowels, and brain." The first and fourth are both living and well ; the first one is a

well-developed five-year-old of quite healthy appearance, with good physiognomy and teeth.

Since marriage the patient has always lived in a dry and comfortable house, but has had at times difficulty in getting sufficient nourishment. Before marriage, and for some time afterwards, she worked at rule-making.

Family history.—Father and mother alive and well, except that the latter has a patch of lupus affecting the right side of the nose, right eyelids, and cheek; this has existed for twenty years, and is still active. The father's sister had her backbone curved from childhood, was never able to speak or feed herself, and died at twenty-three. Two maternal uncles died at thirty-three and forty-four respectively.

The patient is the fourth of eighteen children, of whom the first six and the last one grew up; all the others died at from nine to eighteen months, many of them in convulsions. The eldest brother is "afflicted"; notes of an examination of him are appended to this paper (since the notes were taken, the man has on one occasion tried to commit suicide by hanging himself). The third brother committed suicide. The second, fifth, and sixth, and the youngest of the family, three males and one female, are living, and are said to be healthy and strong.

Present state.—Mrs. A— is 4 feet 7½ inches in height, of fair general muscular development, but with little subcutaneous fat. A scar on the front of the left thigh is said to be the result of a burn at the age of eight.

The patient can stand without support, but has to hold on to something to steady herself in walking; the chief difficulty in walking is at the hip-joint (Plates I and II).

While standing, there is a deep hollow over the lower lumbar and upper sacral regions; the increased curve, with its concavity looking upwards and backwards, affects the whole of the lumbar and the upper sacral vertebræ. There is also some lateral curvature of the dorsal spine, with the convexity to the right. Viewed laterally, the buttocks project about five inches behind the

Adlard, imp.

Adlard, imp.

Adlard, inf.

Adlard, imp.

vertical level of the shoulders, and six inches behind the upper lumbar spine. The abdominal wall sweeps downwards and backwards in its lower half, forming almost a semicircle from the epigastrium to the pubes, which latter is displaced backwards. The iliac crests run downwards and forwards from the posterior superior spines, and their anterior two thirds are nearly vertical (Plate III).

The right anterior superior iliac spine presents a small depression between two prominent points of bone, as if a small piece had been broken off. The great trochanters form prominent projections, extending upwards on either side to within an inch of the level of the highest part of the iliac crest, and an inch higher than the posterior surface of the upper part of the sacrum, which is horizontal (Plate IV).

When the woman sits down or lies on her back, the lordosis above described completely disappears. As the patient lies on her back, the length of the legs is measured from the anterior superior spine to the tip of the internal malleolus. The right leg measures $29\frac{1}{2}$ inches, and is increased by a moderately strong pull to 31 inches; the left one is 30 inches at rest, and 31 inches when extended.

Joints.—There is much grating in both hip-joints, and passive mobility is free, the legs being able to be lengthened by one or two inches, but immediately becoming shortened again on being left free; both grating and mobility are better marked in the right joint. There is no pain on manipulation. Both knee-joints are distinctly loose, and allow a small amount of abnormal mobility; this is more marked in the left. Nothing abnormal is observed in the ankles, and there is no deformity of the chest or upper limbs.

When the patient stands lightly touching the back of a chair, some jerky, irregular movements are observed affecting the arms and trunk. There is marked unsteadiness on standing with the eyes shut.

Lying on the back, the woman cannot raise her legs from

the couch, and when the legs are crossed she has to lift the upper one with her hands in order to get them uncrossed. The knees can be raised fairly well, the feet remaining on the couch. All the movements of both hips, except abduction, are fairly performed; abduction is slight, being interfered with by the position of the upper ends of the femora. The movements at both knees and ankles, and of the joints of both feet, are all well performed.

There is no special wasting of the lower limbs; no rigidity of muscles.

The knee-jerks are both absent; there is no ankle-clonus.

Sensation.—There is deficiency in both tactile and painful sensation in both feet, particularly over the soles. Thermal sensation is tested with glass tubes containing hot and cold water; above the knees these are correctly discriminated, but the patient is usually wrong in her answers about touches with them on the skin below the level of the knees. When the eyes are closed, and the legs passively moved about, and the woman is then asked to describe the position in which the limb is placed, she is always mistaken; this statement applies to both legs, and to the position of knees, ankles, feet, and toes.

The feet are often cold, but sometimes hot and burning. There are severe pains in various parts of the body, sometimes occurring on two or three days in one week, sometimes not for two or three weeks; they are less frequent than formerly. The most common situation is in the arms, beginning about the shoulders and running down to the fingers; the arms "go dead" until the pains are gone, the woman being unable to raise them. The head is also frequently affected, always on the right side of the vertex and above the right ear. Less often now than formerly, pain occurs down the legs along the front of the bones and in the calves. Occasionally there are lower abdominal and inguinal pains.

There is no plantar reflex on either side; the abdominal

and epigastric reflexes are present, more marked on the right side. Sexual desire has never been excessive; for the last few months both desire and gratification have been somewhat diminished. There has been no difficulty in micturition since the bladder was emptied by catheter and the uterus reduced into the abdominal cavity.

Examination of the face, tongue, arms, and hands, reveals no abnormal physical signs. The pupils are below the medium size, do not react to light, contract sluggishly with accommodation, do not dilate on pinching the skin of the neck. The movements of the eyeballs are good in all directions. The ophthalmoscope reveals nothing abnormal. The corneæ are clear. The teeth are good; particularly the incisors are well formed. The patient displays more than the average intelligence of women in her station in life; her memory is very good.

Genito-urinary.—The catamenia have been absent since January 2nd. The bladder can now be emptied voluntarily. The uterus, which is of about the size usual at the third month of pregnancy, is now entirely above the pelvic brim, and is felt in the lower abdomen to the right of the median line, and extending upwards as high as the umbilicus. *Per vaginam* the os is felt high up, the cervix partly softened. The sacrum appears to have its normal curve, and there is no lateral contraction of the pelvic cavity.

Inclination of the pelvic brim.—When the patient lies on the left side, the line joining the promontory of the sacrum with the upper border of the pubic symphysis appears to form an angle of about 10° *in front of* the long axis of the body. On standing, the inclination of the brim is estimated by placing the tip of the finger on the promontory and the thumb on the top of the symphysis; the line joining these two points extends downwards and a little backwards, making an angle of nearly 10° *behind* the vertical axis of the body.

Measurements of the pelvis.—The diagonal conjugate diameter is easily measured by two fingers, and the

average of several trials gives $4\frac{1}{2}$ inches. Dist. sp. il., 10 inches; dist. cr. il., 11 inches; dist. tub. isch., 4 inches.

Examination of the cardiac, respiratory, and renal systems discloses no abnormal physical signs.

Progress of the case.—On April 3rd the patient reported that there had been some dirty-reddish discharge for four or five days; the cervix was somewhat patulous. The red discharge continued, and on May 13th miscarriage took place. I am indebted to Dr. J. J. Kerr for the information that the woman was delivered of a five months' fœtus with membranes and placenta intact, after having strong labour pains for two days; the delivery was natural, and recovery took place naturally.

On July 17th I again saw Mrs. A—, and found her general appearance decidedly improved. The uterus was of natural size and mobility, there was nothing abnormal disclosed on pelvic examination. The condition of the nervous system was carefully gone over, and found to agree in every detail with the notes previously taken and above recorded.

The patient was next seen on February 26th, 1895, when she was found to be again nearly two months pregnant. After this visit she was lost sight of, and was not found until February 28th, 1896, when she said that she had had to go into the Workhouse Infirmary to be confined. The delivery took place on September 18th; for a fortnight previously there were "forcing" pains, which continued much the same in character till the child was born; forceps were applied. There was no trouble with the after-birth. No difference was noticed by the patient in the character of the pains of the second stage between this and former labours. The child is living, strong, and well; it was suckled for a time, but the mother's milk began to diminish at three weeks, and finally disappeared at the end of two months.

For two months before the confinement the woman says she could not stand or walk, nor even sit up; on several occasions she fell off a chair. The inability to

stand or walk continued for two months after the labour, and then rapidly disappeared.

On this date, February 28th, 1896, the condition of the patient was again carefully gone over. The pains seemed to have become, on the whole, less marked and severe than they were. The ability to walk and stand, and the power of movement in the legs, were the same as before. Sensation of all kinds was about the same, except that there was now deferred sensation of the prick of a pin on the soles of both feet. Micturition is normal, except that a call has to be promptly obeyed; frequently a motion is passed unawares, on these occasions there is no warning sensation, and if there were the patient does not think she could control the action. Sexual desire and power are about as they have always been. The pupils are small, 2.5 mm., equal; reactions as before; optic discs normal. Memory and intelligence are perfectly good.

Locomotor ataxy is said by Dr. Gowers, in his 'Manual of the Diseases of the Nervous System,' to occur ten times more frequently in the male than in the female sex; and Erb in one series collected 350 male to only nineteen female cases of the disease. Marie is of opinion that probably 4 to 5 per cent. of cases of tabes suffer from arthropathy, and the joint affection might therefore be expected to be very rare in women. This, however, does not appear to be the case, for in a number of cases of tabetic arthropathy Weizsäcker found seventy-two males and thirty-nine females; although this probably represents too high a proportion of females, it seems to indicate that among patients affected with tabes, females are more liable than males to have the joints affected.

Tabes rarely begins below the age of twenty, and not very commonly under thirty. In the case under consideration the disease began with lightning pains in the lower limbs at the age of sixteen, and the joint affection began at twenty-four during the second pregnancy, dislocation of both hips occurring at the age of twenty-six.

Since this time two pregnancies have gone on to term, and one has ended in miscarriage at the fifth month. Such a case is certainly very rare, and I have been unable to meet with the record of a similar one.

The clinical picture is unusually full of detail: unsteadiness on standing with the eyes closed, athetoid movements, loss of knee-jerks, affection of tactile, painful, and thermal sensation, loss of muscular sense, spontaneous fracture of the ilium, disease of both knees and both hip-joints, temporary strabismus, Argyll-Robertson pupils, pains of various kinds, paræsthesiæ, and some affection of the sphincters,—all were observed.

As regards the ætiology, acquired syphilis, which is a factor in the previous history of from 58 to 88 per cent. of patients suffering from tabes, can, I think, be excluded from the present case with confidence. There are, besides, no signs of congenital syphilis either in the patient or in the brother who was examined. On the other hand, there is a strong family predisposition to nerve degeneration, as evidenced by the paralysis in a paternal aunt, the suicide of one brother, and the attempted suicide of a second.

In four other cases of tabes in females of which I have notes, in two there was a strong presumptive evidence of acquired syphilis; in one, the ataxic symptoms began at twenty-one, and between the ages of sixteen and thirty-five there were nine miscarriages, all at the third or fourth month: in the second there were three children stillborn at term, and one miscarriage at the third month; a sister of the patient suffered from fits.

No clear effect on the progress of the nervous affection in Mrs. A— can be attributed to the repeated pregnancies. The symptoms of tabes seem to have increased rather rapidly from the time of the second delivery onwards for about two years; after this they again became stationary. No material difference in the symptoms and signs has been detected by careful and repeated examinations in the two years during which the patient has been under my observation.

From the obstetric point of view the case is interesting as showing the effect of simple increased inclination of the pelvic brim on pregnancy and labour. The lordosis, and with it the increased inclination, began in 1889, when the woman was twenty-four years of age; the pelvis would then have quite or nearly reached its full development. At the present time, taking the promontory of the sacrum as a centre, the plane of the pelvic brim has rotated downwards through an angle of somewhere about 40° , so that in the erect position the plane of the brim looks forwards and a little downwards. Measurement shows that we have to do with a flattened pelvis of average transverse size and development; the true conjugate is about half an inch below the average. This flattening has given rise to no difficulty which has called for interference during delivery until the last occasion, when labour was completed by forceps.

The rotation of the brim is due in the first place to the lordosis secondary to dislocation of the hips, and in the second to rotation backwards of the anterior or lower half of the os innominatum by the ilio-femoral ligaments, and by the action of the muscles which are attached by one extremity to the ischium and pubes, and by the other to the upper part of the femur. The muscles which will have the greatest effect are the internal and external obturators, the quadratus femoris, the gemelli, and the upper part of the adductors, especially the adductor brevis. The pectineus, the pyriformis, and the psoas and iliacus, will aid the rotation to a less extent.

The lordosis, and with it the increased inclination of the pelvic brim, disappears when the patient sits or lies down; this is a proof that the increased curve is not due to weakness of the spinal muscles, but to the dislocation of the hip-joints, and the consequent lessening of the effect of the extensors of those joints. It has been pointed out by Dr. Gowers that the lordosis which is frequently met with as an early symptom in pseudo-hypertrophic paralysis also disappears on sitting, and is thus proved to

depend on weakness of the extensors of the hip and not of the trunk muscles.

The influence of the alteration in the pelvic inclination was shown in the fifth pregnancy by the uterus becoming incarcerated in the pelvis at the tenth week ; this was the state of affairs when the patient first came under my observation. In the sixth pregnancy the womb rose into the abdominal cavity without any trouble. After entering the abdomen the uterus lay entirely above the brim, so that at the third month it already extended as high as the umbilicus, a height which under ordinary circumstances it only attains at the fifth month. The uterus was displaced entirely to the right of the middle line by the prominence of the vertebral column.

On labour the alteration in the pelvis seems to have had no appreciable effect. The increased inclination of the brim would tend to prevent the head entering ; it was observed, however, that the lordosis, and with it the alteration in the inclination of the pelvis, disappeared when the woman assumed the recumbent posture.

On the pains of labour, the disease of the nervous system, in spite of the impairment of sensibility associated with it, appears to have had no influence. Tabes does not, as is well known, necessarily affect the visceral nervous system, although it may do so ; as Dr. Gowers has pointed out, a fatal pleurisy may be painless.

My thanks are due to Sir Willoughby Wade and to Mr. Gilbert Barling for allowing me to make use of the notes taken when the patient was under their care in hospital ; and I have also to express my sense of obligation to Mr. Bower, late house surgeon at the General Hospital, Birmingham, for his kindness in photographing the patient.

APPENDIX.

G. D—, the eldest brother of Mrs. A—, was seen and examined on March 18th, 1895. His age is thirty-four, height a little over 5 feet; appearance that of a partial idiot.

Speech is very defective, the man being often unable to find words to express himself, and pronouncing those he uses very thickly and imperfectly. He seems to understand well what is said to him in the course of the examination, obeys orders intelligently, and, in fact, investigation shows that he is possessed of a much higher intelligence than would at first sight be supposed. He is said to be able without inconvenience to do without sleep for three or four nights at a stretch, and he often sits awake by the fire all night. His appetite is said to be very large.

The face is decidedly lacking in expression, smooth, the natural furrows few and badly marked. The action of all the muscles on both sides of the face is very defective. Frowning is done slightly, but very imperfectly; the forehead cannot be raised as in surprise; the man cannot grin to order, the left angle of the mouth moving only slightly, the right not at all; both angles move when the patient is made to laugh; the patient can whistle. The tongue is protruded a little to the right. The eyes move fairly in all directions; there is no nystagmus; the pupils react briskly both to light and accommodation. The ophthalmoscope shows nothing abnormal. The upper limbs are normal in power and movements. No radius or triceps jerk can be obtained on either side. In walking the man projects his body somewhat forward, keeps his legs bent at the knees and the toes pointed, and progresses with short quick steps. The leg muscles are well developed; they offer considerable involuntary resistance to passive movements. The knee-jerks are very active, equal on the two sides. There are well-marked clonic movements at the ankles, while the feet retain a partly extended position against pressure, but there is no true ankle-clonus. There

is no defect in tactile or painful sensation anywhere in the body. The plantar reflexes are very active, equal on the two sides. The cremasteric, abdominal, and epigastric reflexes are equal. Sometimes, perhaps once in two or three days, there is involuntary micturition. The bowels are regular, and never act involuntarily.

The condition above described dates from infancy, and is said to have come on with "fits." The brother-in-law who accompanies the man says the disease has been at a standstill ever since he has known him, that is for at least ten years. The patient earns some money by selling papers in the street, and seems to be fairly good at this particular line of business.

Since these notes were taken the man has on one occasion tried to hang himself.

The affection of the nervous system in this case seems to be most likely due to meningeal hæmorrhage from injury during birth, resulting in degeneration of certain of the convolutions, notably those having to do with the movements of the face, tongue, speech, and legs.

(For report of the discussion on this paper, see 'Proceedings of the Royal Medical and Chirurgical Society,' Third Series, vol. ix, p. 9.)