CLINICAL LECTURE

ON

CASES OF SPINA BIFIDA, WITH PARALYSIS OF SPHINCTERS.

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GENTLEMEN,—I have recently had under observation two examples of that rare form of spina bifida in which the patient seems likely to live to adult age, and with no nerve-defect excepting that of double sphincter-paralysis. As you are aware, in not a few cases of spina bifida there is considerable defect in the innervation of the lower extremities, and some form of paralytic talipes (especially calcaneus) may be present. In the cases to which, however, I now refer, the legs are in good condition and the feet straight, and incontinence of urine and fæces is the single, though it must be admitted sufficiently afflictive, consequence. The difference, of course, depends upon the precise nerve-trunks which are involved. You may have large spina bifida tumours in which no nerves are involved, and no paralytic defect whatever is present; and, even when nerve-trunks pass out of the spinal canal into the sac, it by no means follows as a matter of necessity that they will be disorganised. In the present instances, however, we may assume that the fourth sacral nerves (with possibly the fifth and sixth also) pass into the sac and are united with it in such a manner that, for practical purposes, they are disorganised. The fourth sacral supplies a branch to the sacral plexus, whilst its lower half passes directly to the anal muscles. From the lowest strand of the sacral plexus is given off the pudic nerve, which supplies, amongst other structures, the sphincter of the urethra. Thus we see how it comes about that defect of one nerve may involve both these outlets, and lead to the double incontinence of which it is difficult to say which is the more distressful. My patients, as it happens, are of nearly the same age, but of opposite sexes; and it is of interest to remark that sex makes no difference, and that the incontinence is as marked in the one as the other. The muscular sphincter of the urethra is, indeed, precisely the same in woman as in man. I find in the minds of students some little vagueness as to what constitutes the true urethral sphincter; and one of you told me the other day that he supposed the prostate with its muscular tissue served that function. This was, of course, an error. In the long male urethra, several different conditions and structures may combine to make the escape of urine difficult; but there is one real sphincter which it is important to keep in mind. This is the muscle known as Guthrie's muscle, or the *compressor urethræ*, placed between the layers of the deep fascia, and aided by a ring of muscular fibres surrounding the urethra within it. It is these muscles which cause spasmodic stricture; it is these which, when paralysed, permit incontinence. They are supplied, let me repeat, by the pudic branch of the sacral plexus, which is probably derived from the fourth sacral.

I must now proceed with my cases. One of them—the girl—I had an opportunity of showing to you a week ago. She is a well-grown child of about eleven years. I was told that she had a third or superfluous mamma over the nates; and, curiously enough, there is on one part of the cyst a little nipple-like projection of skin. The tumour is about the size of a girl's breast, and looks and feels not unlike one. It is not in the least pedunculated, and is covered at the sides by thick deposit of fat. On more careful examination, you see that it has scars over it, and you find that in its middle there is certainly a quantity of fluid. It is not placed exactly in the middle line, but a little to the left; and there are two depressions in its surface, about which coarse hairs grow. These conditions are not uncommon over spina bifida cysts. It was, of course, present at birth. It is placed over the lowest part of the lumbar region, but it is impossible to say precisely at what part it communicates with the spinal canal. As the girl was being placed on the table, I noticed that she was wet with urine; and, having been told that she had a superfluous mamma in the lumbar region, I guessed at once what was the matter with her. It was only a week before that I had seen a precisely parallel case. We found the child's labia excoriated and eczematous by the constant escape of urine; and, on introducing two fingers into the rectum, we found not the slightest resistance. The opening might be stretched with the

fingers until we could easily look up into the cavity. Thus I infer paralysis of both the external and deep sphincters. I could not appreciate any muscular resistance whatever. We carefully tried to ascertain whether sensation was present, but it was difficult to be certain. It was not wholly lost, but I think it was probably defective. The same condition of easy dilatability existed in the urethra. The girl was reported to walk well, and made no complaint of her feet; but I observed that they were very chilly and blue, although the day was not cold, and they looked much like those of infantile paralysis. Still, I repeat, there was no proven defect of sensation or motion.

The case of the boy to which I have referred was in most respects precisely similar to that of the girl whom you have seen. He was brought by his parents to my house a fortnight ago, the question being as to whether anything more could be done to relieve his condition. He had for long been in the habit of wearing an India-rubber urinal, and his prepuce was swollen and excoriated from the irritation. In him, the spina bifida tumour was smaller than the girl's—not bigger, indeed, than a duck's egg; and it was placed in the middle line over the last lumbar vertebra. He had not the slightest defect in his lower extremities, and was of active habits. His head was, however, large, showing that in infancy there had been a tendency to hydrocephalus. This simultaneous development of spina bifida with hydrocephalus is frequent, and of much interest as supporting an opinion which I formerly advanced, that the former is usually due to intra-uterine dropsy of the meninges of the cord, which prevents ossification.

In reference to the very important question in these two cases, as to whether anything more could be done for their relief, I may admit that I was much at a loss. The incontinence of fæces did not occur all day long, but usually only once or twice a day, or oftener when the bowels were relaxed; and the same was in both cases noticed with the urine. In the girl, I passed a catheter and found a large quantity in the bladder. Still, as regards both viscera, there was entire absence of voluntary control and constant risk of an accident. I advised that the children should be instructed to empty the bladder at regular and short intervals, and invariably before going into company; and that the bowels should be washed out every morning by means of an enema. The father of the boy told me that he thought he experienced some difficulty in expelling urine, for he showed the greatest unwillingness to make an effort to empty the bladder. We can easily understand that it is very possible that the detrusor is weak. In the girl, the friends seemed content with the use of a diaper, and did not speak with any great urgency of the inconvenience. In the boy's case, much more complaint was made.

I do not like to miss this occasion for still further enforcing the physiological lesson of these two cases, that the nerve-supply for the two important sphincters comes from the same source, and that the two may be paralysed together without implication of any other part. I do not know that I ever met with this precise condition and no other complication in any other kind of disease or injury. Yet there are other well-known facts which depend for their explanation upon the same anatomical distribution of nerves. When a desire to defecate is present, you do not dare to attempt to relieve the bladder excepting at the closet, for fear that, in relaxing the sphincter urethræ, you will let go also that of the bowel. If a patient be suffering from spasmodic retention of urine, there is no better plan than to give him a good dose of croton-oil or Epsom salts; for, when the sphincter ani and associated muscles are made to relax beyond the slightest possibility of restraint, it is ten to one if the sphincter urethræ can maintain its spasm. You may by this means obtain valuable aid to diagnosis; for, if retention of urine persist after free purgation, it is highly probable that it is due to something more than spasm.

I have not discussed any question of the treatment of spina bifida in these cases. My opinion is, indeed, so definite on this point, that I did not for a moment think of attempting anything more than relief of the incontinence. Nothing whatever would have been gained as regards the incontinence by the removal of the tumour even had it been safe to effect it. As regards getting rid of the tumour itself, there was really no object whatever to be served by attempting it. The tumour was not in either case in the least in the way. Were it not for the incontinence of fæces and urine, neither patient would have cared in the least about the abnormality. It was for these symptoms only that relief was sought. Now, excision of the cyst, or its obliteration, could not possibly affect the nerves contained in it otherwise than injuriously. It might make their state worse, but could not better it. I repeat, then, we had no temptation whatever to meddle with the tumour. In infants, sometimes, when the cyst is very large and tense, we may feel much attracted to the proposal to effect its reduction by tapping, or to try to obliterate it; for here its mere bulk is an inconvenience. When the child has lived on to the period of puberty, how-

ever, this motive is always removed, for the tumour is always reduced in size and perfectly quiet as regards increase.

One case is on record in which a surgeon, usually of most excellent judgment, was persuaded by entreaties of friends to do an operation in a girl aged nine. It appears to have been a case exactly like those which I have mentioned, incontinence of urine and fæces being the only symptom. The cyst was protected by a thick covering of fat. This was carefully dissected aside and the cyst opened; and, being found apparently free of nerves, a part of it was excised. The girl died in a few days of meningitis of the cord. It may be added that treatment by puncture and compression had been repeatedly tried before, and without any advantage.

REMARKS

ON

THE PHYSICAL CAUSES OF THE SOUNDS OF THE HEART.

By CHARLES J. B. WILLIAMS, M.D., F.R.S.,

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IN the number of the BRITISH MEDICAL JOURNAL for May 12th is a communication from Dr. Little, charging me with some inconsistencies in my explanation of the sounds of the heart, and complaining that I have not given sufficient credit to Dr. Billing as "the discoverer" of the valvular origin of the sounds of the healthy heart.

I am very sorry if I have not done justice to my venerable friend in this matter, and I have waited till my return to London to enable me to refer to the several works on the subject.

The first published statement of Dr. Billing's views I find in the *Lancet* of May 1832, under the title, "On the Auscultation and Treatment of Affections of the Heart, read at the Hunterian Society in February 1832". In this, Dr. Billing says:—"The first sound is caused by the tension produced in the shutting up of the auriculo-ventricular valves, and the second sound is caused by the tension produced in the shutting of the ventricular-arterial valves." This early and clear statement of the valvular theory of the heart-sounds was not known to me when I wrote the summary of views on the subject, which appeared in the third edition of my little work on *Diseases of the Chest* in 1835 (quoted by Dr. Little), and I regret that, through this ignorance, I did not give Dr. Billing's name its proper place among the advocates of that view. But, in my fourth edition, published in 1840, I mention that "this view was proposed by Dr. Billing in February 1832; but, as far as regards the second sound, he was anticipated by Dr. Carswell, whose opinion was publicly announced in the Academy of Medicine at Paris in July 1831". My authority for this statement was the following note in the *Library of Medicine* by Dr. Joy, vol. iii, p. 262:—"The discovery of this theory of the second sound is claimed by Dr. Carswell, and was first publicly announced by M. d'Espine in a dissertation read by him at the Academy of Medicine at Paris in July 1831, though it was made known by Dr. Carswell as early as 1829. There is in the museum of University College a drawing (dated 1829) of the case of an aneurism of the aorta observed by Dr. Carswell in the hospital of La Charité, the physical signs of which case first suggested to him this now almost universally received theory."

In the thesis of Dr. Elliott (*De Corde*, Edinburgh, July 1831), to which my attention was called by my most revered teacher the late Professor Alison, is found the earliest announcement in print of this explanation of the second sound that I am acquainted with. Dr. Billing denies that Dr. Elliott ascribes the second sound to the arterial valves, because he considers the rush of blood from the auricles at the diastole of the ventricles also concerned in its production; but the words which I put in italics in Dr. Billing's quotation from Dr. Elliott's thesis seem to me plainly to point to the arterial valves as a chief cause of the second sound:—"Naturam soni secundi subitam et abruptam fieri credo a diastole ventriculorum tam repente et vehementer inchoatâ ut sanguis vi magnâ auricularum parietes transcurrat: necnon ab impedimento quod in corpore sano ferè instantè valvulâ præstant sigmoideæ sanguini qui in ventriculos, dum horum fit diastole, ex arteriis vult refluerè."

But, although I thus find reason to question Dr. Billing's priority in proposing the valvular theory of the second sound, I am ready to accord to him the full merit of originality in ascribing the first sound to the tension of the auriculo-ventricular valves, and to admire the zeal and ingenuity with which he has supported that view. But I think that

his zeal for his favourite opinion carries him too far when he denies that muscular contraction has any share in producing the sound, as in the following passage. "I maintain that the first sound as well as the second is entirely valvular, and deny that any part depends on muscular noise; for, when there is simple concentric hypertrophy (increase of muscle and muscular action), the cavity of the ventricles being diminished, there is diminution of sound, although there is more of the condition necessary to the supposed *bruit musculaire*." (*First Principles of Medicine*, 1868, p. 43.)

I have already, in a previous communication (BRITISH MEDICAL JOURNAL, February 3rd, 1877), as well as in former works, refuted this objection to the muscular origin of the first sound, by proving that the sound of muscular contraction is loud in proportion to the thinness and abrupt action of the muscle, and inversely as its thickness. The movement which causes sound is the sudden change from slack to tight, which is greater in thin muscles and in a dilated heart, and less in thick muscles and in an hypertrophied heart. In an appendix to the second edition of my *Rational Exposition of Physical Signs* (published in 1833), the various views of the causes of the sounds of the heart are discussed in order; and, in reference to the first sound being caused by the closing of the auricular valves, I remark: "The principal objection to this as the only cause of the first sound is, that it must be instantaneous and confined to the first part of the ventricular systole, whereas we know that the first sound is prolonged during the whole period of this action" (p. 199). The same consideration had several years before led me to suppose the muscular contraction itself to be the essential cause of the first sound; and further reflection and investigation led me to refer it to a general law, that sound is produced wherever a soft solid is suddenly and forcibly tightened. The forcible transition from slack to tight in muscular action fulfils the conditions of motion and resistance which constitute sound. After the analytic demonstrations which I considered to have been made by my experiments in 1835, I endeavoured to remove the mystery of the *bruit musculaire* by referring both it and the first sound of the heart to the simple principle of sound caused by resisted motion in the tightening or tension of muscular action. If Dr. Little finds any variation of expression in my later writings from that used in 1828 and 1833, it is not from discrepancy, but from the fuller development of the subject, and from my having been able to trace the sounds of the heart to the general fact of movement from slack to tight, including not only the tightening of the muscular walls, which is the primary and most essential part, but also in the tightening of the valves, which is an accessory and auxiliary cause.*

In conclusion, I beg to remind Dr. Little that my first communication, entitled "A Few Personal Facts concerning Experiments on Animals, etc." (BRITISH MEDICAL JOURNAL, July 22nd, 1876), did not profess to claim for my experiments the *discovery of new views*; the experiments were performed to decide between the many views or conjectures previously advanced to explain the sounds of the heart, and the results proved so positive and so conclusive, that they left little doubt in the minds of those present; and, after having been repeated by subsequent experimenters, these results have been generally confirmed. Dr. Billing's view of the exclusively valvular origin of the heart-sounds was, doubtless, brought forward at a very early period and with great ability; but, although rendered probable by various clinical observations, it had not been brought to the test of experimental trial. As far as regards the second sound, my experiments proved the correctness of Dr. Billing's views; but they showed their insufficiency with regard to the first sound, inasmuch as this sound was distinctly heard when the action of the valves was stopped. This positive result is more conclusive than the negative results of Dr. Halford's experiments with the bull-dog forceps on a dog. It is asserted that by strong pressure with this instrument on the afferent veins the heart was deprived of its blood, and all sounds ceased, although the ventricles continued to act. As I have before stated (BRITISH MEDICAL JOURNAL, September 30th, 1876), I question the accuracy of this observation as to the complete extinction of the first sound; but I should expect it to be much weakened, not only from the feeble action of the heart deprived of its natural stimulus, but also from the absence of contents to spread the walls into a vibratile condition. An empty sac contract-

* Dr. Little criticises my use of the word "flapping", as applied to the tightening of the valves, because he thinks that the word implies something loose, like the sail of a ship flapping with the wind. I confess that I first used the term in an onomatopoeial sense, like the words clap, rap, slap, etc., as signifying a motion making an abrupt sharp noise; but I am ready to vindicate my use of the term even "at sea" with Dr. Little, by proving that a loose sail flaps noisily only as it is blown tight by the wind from or against the mast or rigging. The heart-valves do not "flap about" in the blood, as the loose sail does in the wind, irregularly; but they do flap, or pass from loose to tight with a noise, in a definite manner, in obedience to the motions of the heart and the pressure of the blood.