

TWO CASES OF MALFORMATION OF THE KIDNEY IN INFANCY.

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THE following is a brief record of two interesting cases of the above nature which have appeared in our hospital practice during the past two years.

CASE 1.—B. L., aged 14 months, a full-time baby weighing 8 lb. at birth. Congenital malformation of the ureters and bilateral hydronephrosis, upon which was superimposed a *B. coli* infection. Admitted to hospital with history of recurrent vomiting and wasting since birth. Every kind of diet had been tried without effecting any definite improvement in the general condition. Picture, as presented on admission, of obstinate infantile dyspepsia; the baby was fretful, the abdomen distended and hard; stools pale, undigested, and sour-smelling. The urine slightly offensive; marked emaciation, the weight being only 14 lb. 9 oz. A fortnight before admission a small quantity of bright red blood had been passed per rectum on one or two occasions.

On Examination.—Abdomen difficult to palpate, owing to its abnormal distension and hardness; in left iliac fossa an elongated sausage-shaped tumour, which suggested at first sight thickened pelvic colon or a chronic intussusception. The urine contained a quantity of pus cells, and on cultivation a growth of *B. coli* was obtained. The pulse, temperature, heart, lungs, and other systems were normal. A few days after admission typical *B. coli* rigors took place, and a moderate degree of temperature persisted despite treatment until death. So far the case, with the exception of the indefinite tumour in the left iliac fossa, had appeared to be one of defective digestion and mal-assimilation associated with a *B. coli* pyelitis. Five weeks after admission, however, the infant began to vomit all feeds, became dyspnoeic and drowsy, and very shortly passed into coma and died.

Post Mortem.—A condition of bilateral hydronephrosis was shown, with marked dilatation and convolution of the ureters, which were duplicated on either side. The elongated tumour felt during life was an enormously dilated and tortuous left ureter. The kidneys, ureters, and entire bladder were removed and submitted to a further dissection, an account of which follows:—

The kidneys were enlarged, the left being $3\frac{1}{2}$ in. long, and the right $3\frac{1}{4}$ in. The hilum of the left organ was large, and emerging from it were two pelves which were prolonged downwards as two separate ureters; the latter formed wide thin-walled tubes, pursuing a tortuous course to the back of the bladder. Just outside the bladder wall the ureter which sprang from the lower half of the kidney tapered away to a narrow vessel and entered the dilated end of the other ureter; the single tube then pierced the bladder wall and opened at the summit of a rounded cyst-like body which projected into the vesical cavity on the left half of the trigone. This opening was in the form of a small slit about one-eighth of an inch long. Fluid could be expressed from both ureters through these small openings. Viewed from above the pelves were dilated and the calyces greatly enlarged; the lining membrane was smooth, while a thin layer of kidney tissue formed the capsule of this large collection of cyst-like cavities. The upper pelvis and its calyces were separated from the lower ones by a thin septum of kidney tissue. The right kidney presented an exactly similar condition to the left, except that the ureters were rather smaller and less convoluted. The openings of the ureters on the cyst-like prominences in the bladder were valvular, the lower lip of the orifice projecting beneath the upper, so that pressure within the sacs would cause the lower wall of the opening to bulge upwards and obstruct the flow of urine into the bladder. The bladder itself was a little enlarged and the wall thickened, especially at the fundus. The urethra was wide and unobstructed, but the right ureteral sac tended to bulge over the orifice. The wall of the urethra itself was thickened.

CASE 2.—This case is somewhat similar. L. B., a female infant, aged 7 weeks, was brought to hospital for an apparently mild attack of diarrhoea and vomiting, was treated and referred to out-patients, and on the following day died quite suddenly.

Post Mortem.—A condition of widely spread oedema of the whole body was shown. On opening the abdomen both kidneys were found to be slightly enlarged, both exhibiting a moderate degree of hydronephrosis, the pelves and calyces being widely dilated and the kidney tissue reduced in each instance to a narrow belt one-quarter to one-third of an inch in diameter. From the upper pole of the right kidney there arose from a narrow orifice a widely dilated and convoluted ureter an inch in diameter. This ureter ended blindly in the right side of the bladder wall. From the pelvis of the same kidney a moderately dilated and tortuous ureter descended and opened normally into the bladder.

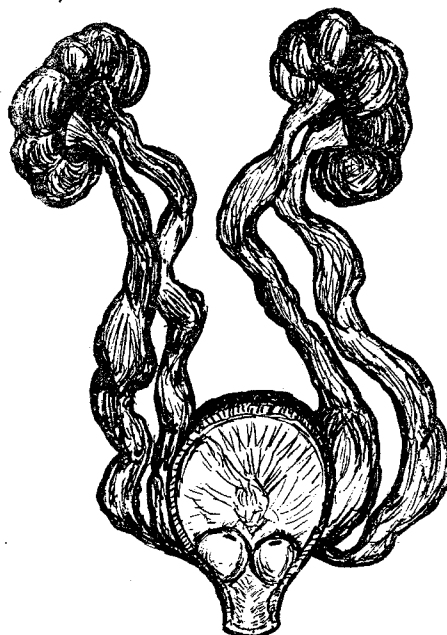


Diagram to illustrate Case 1.

From the left kidney a similar malformed ureter arose normally from the pelvis and presented a normal ureteric orifice in the bladder. The wall of the latter was distinctly thickened and hypertrophied. No signs of obstruction could be detected at either opening of the ureters or at the urethral orifice. Sections of the kidney tissue showed no abnormal changes.

Remarks.

It is not easy to explain the pathological changes observed in these two cases. There is a congenital malformation present, but the causation of the dilatation of the ureters and hydronephrosis is not so apparent. At first sight it would appear that the obstruction must lie at the ureteric orifice, but in both cases, with the exception of the ureter which ended blindly, the openings were large enough to allow of a free passage of the urine. In the first case, with its cyst-like prolapse of the ureters into the bladder, the valvular nature of the orifice caused by the prolapse may have been sufficient to check the flow of urine. But it must be remembered that this prolapse was itself secondary to either obstruction at the ureteric orifice or lower still. Again, in both cases the bladder was thickened, indicating the possibility of some obstruction to the out-flow through the urethra. In both cases, however, the patient was a female, and the short urethra was widely patent, and there was no sign of the obstructing flap of mucous membrane described by some writers. Torsion of the greatly convoluted ureters hardly seems a satisfactory cause, and the possibility of congenital hypertrophy of the muscles of the bladder compressing the ureteric orifices seems little better. If none of these causes are operating we are left to presume that there must be some incoördination of muscular action causing spasm of the urethral orifice. Such a condition would be comparable to the spasm seen in pyloric stenosis or in the obstinate constipation resulting in or from Hirschsprung's disease—i.e., congenital dilatation of the colon.

Since writing these notes our attention has been drawn to a paper by Dr. John Thomson,¹ in which he classifies this condition with congenital pyloric hypertrophy and hypertrophy of the colon.

¹ Brit. Med. Jour., 1902, ii., 678.