THE CLINICAL DIAGNOSIS OF CONGENITAL ANOMALY IN THE KIDNEY AND URETER.

BY WILLIAM F. BRAASCH, M.D.,

OF ROCHESTER, MINNESOTA,

Attending Physician to the Mayo Clinic, and St. Mary's Hospital.

IN the literature of the past few years one may frequently find reports of one or two cases of congenital anomaly in the kidney and ureter. The existence of this condition was usually discovered at operation or at autopsy, and no extensive reference was made in regard to the exact clinical diagnosis. With the development of the cystoscope and the radiograph and, more recently, through the discovery of the value of these instruments in their simultaneous employment, as in pyelography,[1] we now have at our command the means to make an accurate diagnosis in practically every case of renal or ureteral anomaly.

The frequent occurrence of congenital anomaly in the kidney and ureter is not generally appreciated. A review of the surgical and clinical records of the Mayo Clinic for the past five years shows that gross renal and ureteral anomalies were found in 36 patients. Of this number, seven were operated for diseased conditions in the abdomen other than those in the kidney, in whom the discovery of the renal anomaly was largely incidental to general abdominal exploration. Eighteen were operated for various pathologic conditions complicating the anomaly. In the last three years we have been able to make the clinical diagnosis of congenital anomaly in 14 patients; of this number, four were not operated for various reasons. Post-mortem records of the last 171 autopsies made at the clinic showed that congenital anomaly of the kidney and ureter was noted in seven cases or over 4 per cent. of the total. This number includes only those gross anomalies occurring in the adult that might be regarded of surgical importance, and does not include minor anomalies so frequently found in

the urinary tract, such as supernumerary and aberrant renal blood-vessels, fetal lobulation, moderate degree of malposition, nor those rather frequent instances of partially deformed or moderately atrophic kidneys which may be due either to congenital or acquired etiologic factors.

The various anomalies in the order of their frequency were as follows: fused or horseshoe kidney, 11; congenital, single, or asymmetrical kidney, 6; atrophic kidney, 5; ectopic kidney, 3; duplication of renal pelvis and ureter, 8; division of ureter, 5.

The surgical records of the Mayo Clinic show 649 operations on the kidney and ureter during the past five years. Excluding the four cases diagnosed and not operated and the seven cases found at autopsy, there remain 25 cases operated, or a proportion of 1 congenital anomaly to 26.

The pathologic condition existing in the anomalous kidney or ureter usually calls our attention, clinically, to its existence. That such kidneys are peculiarly liable to disease has been noted by various observers. The frequency with which such anomaly is found in a surgical clinic as compared with post-mortem records of a general hospital would, therefore, be at least partially explained by the fact that the complicating conditions usually require surgical treatment.

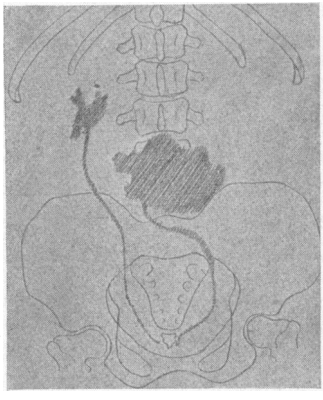
FUSED KIDNEY.

The fused kidney may assume any of a great variety of forms and may be situated in various parts of the abdomen. The type of fused kidney more frequently found is the so-called horseshoe kidney. Although its usual position is in the median abdomen at about the level of the umbilicus, it often lies more to either side of the spine. Occasionally, it will be found lying diagonally, with one pole extending down into the bony pelvis. If our attention be called to the existence of a fused kidney clinically, it is usually because of some pathologic process which causes tumor or localizes pain in the median abdomen. Unfortunately, the *subjective symptoms* caused by various pathologic conditions found in the horseshoe kidney might

easily be confused with the symptoms caused by disease in the surrounding organs. When the complication involves one-half of a symmetrical lying horseshoe kidney, the pain may be more lateral than median, as proved to be the case in three of our series. When the kidney lies asymmetrical, the median lying pole, which is usually the lower, is generally the one involved, with consequent distinctly median pain or tumor. Rovsing [2] has recently described three cases of symmetrical lying horseshoe kidney without visible pathologic changes, which, because of their position, evidently caused pressure on adjacent nerve-trunks. He regarded the resulting pain, referred across the abdomen, appearing on exercise and relieved when recumbent, as pathognomonic. Our series includes one such case, but it was quite impossible to make the clinical diagnosis from the subjective symptoms. *Palpation* may determine a mass lying across the lower median abdomen which is suggestive of a fused kidney but certainly would not identify it as such. Furthermore, in a large or tense abdomen it may be quite impossible accurately to determine a small median tumor such as may be caused by a median kidney. A retroperitoneal mass felt through the rectum may offer corroboratory data, particularly if the pulsation of adjacent large blood-vessels can be determined. This finding, however, can be obtained with various retroperitoneal conditions other than those of the kidney (Fig. 1).

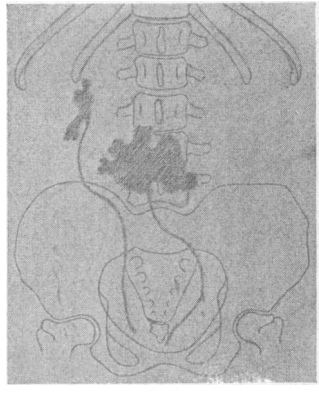
The *radiographic shadow* of soft tissues in the abdomen is usually too inexact to permit of accurate interpretation. Occasionally the outline of a median mass in a thin subject may be suggestive of a horseshoe kidney. Should the radiograph show a shadow of an evident renal stone in the lower median abdomen the possibility of a fused or ectopic kidney would be indicated. Meatoscopy may be of some corroboratory value. In four of the five cases thus examined, the meati were found in their usual position in the trigone. In one, the meatus of the ureter leading to the median lying pole was found in the median line of the trigone. Obstruction to the ureteral catheter at the level of a pelvic mass might be sug-

FIG. 1.



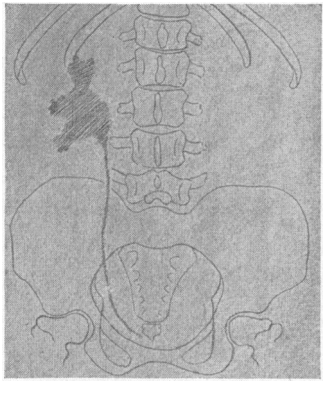
Case No. 10,215. Horseshoe kidney. Hydronephrosis median and lower pelvis. Upper pelvis normal. Kidney resected.

FIG. 2.



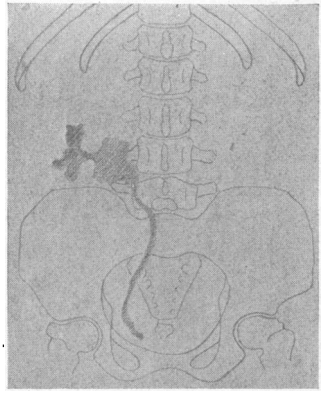
Case No. 9508. Fused kidney. *a*, hydronephrosis median and lower pelvis; *b*, upper pelvis normal. Kidney resected.

FIG 3.



Case No. 5244. Congenital single kidney. Note *a*, large pelvic outline; *b*, normal contour of calyces.

FIG. 4.



Case No. 11,926. Ectopic kidney. Note *a*, peculiar insertion of ureter (pelvis situated anteriorly); *b*, duplication of pelvis.

gestive of some renal anomaly. However, more exact data are to be obtained through the combined use of the radiograph and the cystoscope, namely, through pyelography. The relative position of the pelvis is accurately determined, and any complicating dilatation or deformity of either pelvis and ureter can be clearly demonstrated. Within the past three years we were fortunate enough to secure three pyelographs of fused kidneys, all of which were complicated by a hydronephrosis in one of the poles (Fig. 2).

On reviewing the eleven cases of fused kidney in our series, we find that eight were of the so-called horseshoe type and three were so-called sigmoid kidneys, one with three separate pelves. The various conditions found complicating the anomaly were as follows: hydronephrosis, 4; tumor (sarcoma?), 1; renal hæmaturia (essential), 1; abdominal pain, 1; discovered in operating for other abdominal conditions and evidently otherwise normal, 4. The sex was about equally divided, there being 5 males and 6 females. Six of the patients were below 30 years of age, and of these, five were operated for some complication of the kidney itself. This is of considerable interest and is in keeping with the fact that if complications (particularly certain forms of ureteral constriction) occur as the result of congenital anomaly, they will occur in the young adult, usually soon after their full development. The majority of anomalies occurring in patients over 30 years of age was discovered accidentally at the time of operation for some other abdominal lesion. Evidence of congenital anomaly in other organs was found in but one patient, a female, who was found to possess a bicornate uterus.

CONGENITAL SINGLE KIDNEY (FIG. 3).

The diagnosis of the congenital absence of one kidney can be made clinically only by means of the cystoscope, and thus the condition becomes a problem largely of cystoscopic technic. Naturally, the inability to find a ureteral meatus in a markedly inflamed and contracted bladder does not necessarily indicate its congenital absence. In the hands of an

experienced observer, however, the absence of any evidence of a meatus in a bladder which permits of thorough cystoscopic examination would be strong evidence of a single kidney. To further corroborate this, the segregator, if carefully employed, may be of value. The injection of indigo carmine is often of aid in determining the existence of a meatus. Some difficulty may be encountered in differentiating clinically between acquired and congenital single kidney. When a kidney and ureter become functionless, such as occurs with a so-called autonephrectomy, the site of the former meatus will frequently remain visible and show evidence of some inflammatory change. With the congenital single kidney the meatus is frequently situated in unusual positions, either in the median (one case) or extremely lateral base of the bladder (one case). Close examination of the meatus may reveal an hypertrophy of the muscular ring about it. The peristaltic contraction will be found exaggerated. The secretion may be found unusually frequent or with unusual volume.

Of considerable interest is the pyelograph of the single kidney. The injected pelvis of the hypertrophied congenital single kidney will usually appear considerably enlarged but otherwise quite normal in outline.[2] The increase in the size of the pelvis will be commensurate with the increase in parenchyma. On the other hand, with an acquired single kidney, the several pyelographs I have made do not show any increase in the size of the pelvis.

Although a portion of the other ureter has been reported to exist by various observers,[3] in none of the three cases cystoscoped and operated nor in the two cases reported in the autopsy records was any evidence found of the other ureter.

A quantitative estimate of the functional capacity of the single kidney may be of some corroborative value. The phenolsulphonphthalein test is well adapted for this purpose. A very high percentage of the chemical substance secreted, such as usually occurs with two normal kidneys, might be indicative of the degree of compensatory hypertrophy.

Subjective symptoms are of little value.

Palpation.—To be able to palpate but one kidney and to determine an evident increase in its size is at best only suggestive of a single kidney. It must be remembered that one of two normal kidneys is occasionally found considerably enlarged without apparent reason. Frequently it is quite impossible to differentiate hypertrophy from a large normal kidney lying low and prominent in a thin abdomen. It may be difficult to differentiate between a small renal tumor and an hypertrophy. A tumor of the surrounding organs may closely simulate an enlarged kidney. If, however, on exploration in the abdomen, the surgeon finds that one kidney is unusually large without apparent cause, the other kidney should invariably be searched for, and if present carefully examined for evidence of disease.

Of the six cases of congenital single kidney in our series, four were diagnosed clinically and demonstrated at operation. In one case the single kidney was tuberculous and in another the ureter was kinked so as to cause a moderate degree of mechanical obstruction. In but one patient was there any co-existing anomaly, namely, a bicornate uterus, a congenital absence of cervix, vagina, left tube, and ovary. The kidney was found markedly hypertrophied in every instance. This leads one to infer that where no marked hypertrophy is noted, failure to find the other kidney may not necessarily be a congenital defect.

ATROPHIC KIDNEY.

Atrophy of the kidney can be either congenital or acquired, and it may be quite impossible to differentiate the etiologic factors on gross examination. With a marked degree of atrophy of one kidney the other kidney is usually found hypertrophied. The discovery of hypertrophy in a kidney on abdominal exploration would necessitate examination of the other side. The clinical diagnosis of an atrophic kidney may be exceedingly difficult, and when made is largely dependent on systematic data.

A moderate degree of atrophy could easily be overlooked

in a cystoscopic examination. An evident diminution in amount of secretion on meatoscopy could easily be explained by reflex inhibition of secretion so frequently seen as the result of cystoscopic irritation. A qualitative estimate of the functional activity as evidenced by the length of time required for the kidney to secrete a chemical substance previously injected subcutaneously would be of little value so long as any normal kidney tissue is present. By means of a quantitative estimate we should be able to ascertain the functional capacity more accurately than by any other means. Unfortunately, however, with a diseased kidney on the other side the vagaries in functional estimate in the supposedly normal kidney are often so great that even a marked diminution in functional capacity does not necessarily indicate that the kidney in question is incapable of normal functional activity. Although a moderate degree of atrophy may be present without much evidence on cystoscopy, with marked atrophy, we have various data which should call our attention to its existence. Examination of the meatus will show an atrophy of the circular muscle usually seen about the normal meatus. The meatal contraction will be slight, the secretion will be small in amount and seen but occasionally. The other meatus will usually show a corresponding compensatory increase.

Atrophy of the ureter is usually in keeping with the degree of renal atrophy. This is often the case in conditions of reduced renal secretory tissue in conditions other than congenital. Even a small ureteral catheter may meet with difficulty in introduction as a result of atrophic reduction in size of the ureteral lumen. The pelvis of the atrophic kidney, particularly in the congenital, may be so rudimentary that its outline in the pyelograph may be corroborative of the foregoing data.

The patient's general condition, blood-pressure, ophthalmoscopic data, as well as subjective symptoms, should call our attention to the existence of renal insufficiency in cases where the secretion from an atrophic kidney may appear normal. Atrophy of one kidney to such a degree that it would seem impossible to sustain life, and without apparent cause other

than congenital, was found in five patients. In two cases the condition was found at autopsy and in one case it was demonstrated clinically. The kidneys were described as infantile in size and without other apparent evidence to cause their atrophy. The ureter was found small and atrophic in every case.

ECTOPIC KIDNEY (FIG. 4).

Anomaly in the position of the kidney may be acquired or congenital. A moderate deviation from the normal situation or a freely movable kidney is not necessarily considered a congenital anomaly. When, however, the kidney is found lying fixed in the bony pelvis or across the spine and when its blood-vessels come from adjoining vessels, such as the iliacs, it must be regarded as a true congenital anomaly. Such a kidney is called an ectopic or pelvic kidney. Its clinical diagnosis is easily confused with various conditions in the surrounding organs. Subjective symptoms are usually referred to the lower abdomen and pelvis. On abdominal palpation the ectopic kidney may readily be mistaken for an appendiceal mass or a tumor of the adnexa. On rectal examination it may, in some cases, be felt as a retroperitoneal mass, and when large blood-vessels are felt with it the finding may be of value. However, the only accurate method of establishing the diagnosis clinically is by means of the cystoscope and the radiograph. On cystoscopy an anomalous position of the ureteral meatus may be present. The ureteral catheter can usually be introduced but a short distance. The position of a metal stilette or catheter as shown by the radiograph would localize the position of the kidney, providing the stylet can be introduced into the pelvis. More graphic and complete, however, is the pyelograph, which not alone localizes the position of the kidney but also demonstrates any anatomic peculiarities in the renopelvic outline. It must be remembered that the relative position of the two pelves of an asymmetrical fused kidney, one lying in a normal position and the other lying low in the median line, might simulate the relative position of the pelves of a normal lying and ectopic kidney.

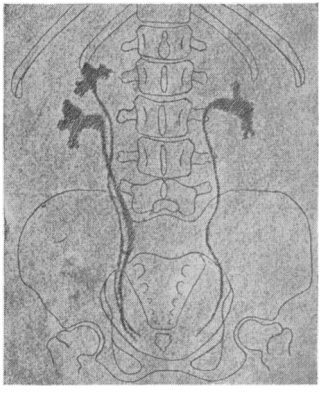
Since it is the existence of some pathologic condition which calls our attention to the anatomic anomaly, it is even more important that we demonstrate this complication clinically. The data obtained through the cystoscope and ureteral catheter alone will aid us materially in ascertaining the identity of the complication. The pyelograph will usually be found to be of greater value, however, to determine accurately the position and extent of any pelvic distention, whether mechanical or inflammatory, and to demonstrate any deformity resulting from inherent tumor. The most common complication associated with the ectopic kidney is hydronephrosis, evidently as a result of its anomalous position.

Ectopic kidney was found in three patients in our series. This number does not include those numerous instances where the kidney was found lying low in the abdomen nor even those freely movable kidneys found in the bony pelvis. It includes only such as were fixed in the bony pelvis and whose blood supply came from the adjoining vessels. As various observers [4] have noted, malformation of the various genital organs frequently occur with such a kidney.

ANOMALIES OF THE URETER (FIG. 5).

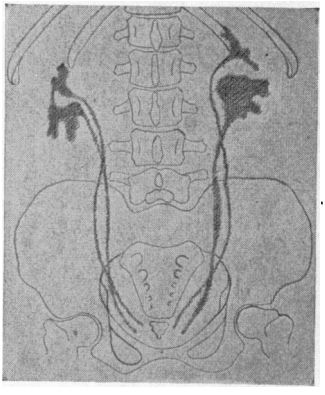
Duplication of the Pelvis.—That the normal renal pelvis may assume any of a great variety of shapes is well known. The individual calyces may be so large and so situated that they resemble separate pelves, particularly so when the calyces do not unite until well beyond the hilum. When, however, there are two distinct pelves within the hilum and each has its separate calyces and ureter, the condition must be considered as an anomalous duplication of the pelvis and becomes of practical importance. Our series includes eight cases of duplicated renal pelvis, five of which were operated upon, one was found at postmortem and two were demonstrated clinically but not operated. In the six cases explored evidence of a division of the two halves of the kidney was externally visible, varying from a slight depression to a distinct area of demarcation. The division is furthermore indicated in cases of true

FIG. 5.



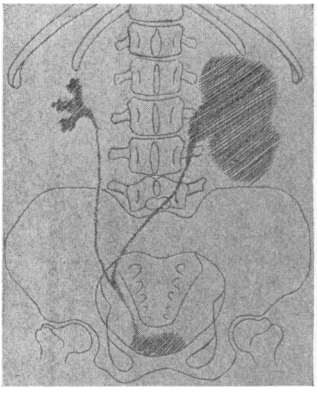
Case No. 10,962. Duplication of the pelvis of right kidney and ureters. Normal. Accidental discovery in routine clinical examination.

FIG. 6.



Case No. 13,529. Bilateral duplication of ureters and pelves. Duplication left kidney pelvis complete with small hydronephrosis and stone of the lower pelvis. The two pelves of the right kidney united by common calyx.

FIG. 7.



Case No. 11,425. Division of ureter at lower third. Left branch crosses spine and leads to a large hydronephrosis of the left kidney. Right kidney normal. Pyelograph obtained by using large catheter and inserting short distance into meatus.

duplication by the fact that the individual poles of the kidney will have in the main a separate blood supply. For practical purposes, therefore, the kidney might well be considered as made up of two distinct kidneys which will permit of separation if necessary. Bisection of such a kidney was performed successfully in three instances by W. J. Mayo after the clinical diagnosis had been made. In one case duplication of the pelvis with separate ureters was found on both sides. In the right kidney the two pelves were united by a narrow calyx in common with both. This connection was suggested by injecting methylene blue solution into one ureter and its return through the other. The separation of the pelves in all other cases was complete. The diagnosis of the pelvis duplication can be made accurately only by means of the cystoscope and radiograph combined. The finding of two separate meati on one side of the base of the bladder on cystoscopy does not necessarily indicate the existence of separate pelves, since the two ureters may unite above the bladder. Neither would two catheters introduced into separate ureters necessarily establish the existence of a duplicated pelvis, since a single pelvis may have two distinct ureters. The pyelograph offers the most accurate means to determine the relative position of the pelves and the amount of renal parenchyma separating them. Furthermore, it will determine the character and extent of any pelvic deformity which may result from some pathologic condition affecting that part of the kidney. The various pathologic conditions affecting the half of the kidney and necessitating operation were as follows: hydronephrosis resulting from anomalous blood-vessels constricting the ureter near the ureteropelvic juncture, 2; hydronephrosis resulting from stricture at the wall of the bladder, 2; hydronephrosis and stone, 1.

Duplication of Ureter (Fig. 6).—In seven of the cases of duplicated pelvis in our series the two ureters were found separate along their entire course and leading into separate meati in the bladder. In one case the two ureters were found lying in close apposition within a distinct sheath for a distance of about 6 or 8 cm. along their middle third, but without any

evidence of anastomosis. The position of the two meati in the four cases cystoscoped and diagnosed was found to be variable. In two cases one meatus was found at about its normal position while the other meatus was situated posterior and more median about 3 cm. apart. In the other two cases the distance separating them was about 1 cm. The stilette or pyelograph will graphically demonstrate the usual crossing of the ureters at the brim of the pelvis. In one case we were able to demonstrate clinically complete duplication of the ureter on both sides, with partial duplication of the pelvis of the right kidney and complete duplication in the left kidney.

Division or Partial Duplication of the Ureter.—The ureter may divide at any part of its course. The most frequent point of division is at the first portion of the ureter where two more branches of the ureter leave the hilum and unite at a short distance below. Often, however, this will not be a true ureteral division but will represent the absence of a true pelvis with the union of extended infundibula instead. Division of the ureter into two branches ending in adjoining meati is occasionally met with. This division may occur along any part of its course, more often in the lower portion. Union of the two ureters arising from normally situated kidneys and merging at about the brim of the pelvis and entering a single meatus on the right base of the bladder was found in one case of our series. This series includes five cases of division of the ureter; one case of double pelvis with two separate ureters joining a short distance below the hilum; one case of three ureters leaving a single large pelvis at various angles of the kidney and uniting at a short distance below, which was found at autopsy; two cases of division of a single ureter at the brim of the pelvis entering separate meati on the same side; one case of union of the two ureters arising from separate kidneys at the brim of the pelvis as described above and which was diagnosed clinically by means of the injected radiograph (Fig. 7).

The clinical diagnosis of duplication of the ureter can usually be made quite easily by means of inserting metallic coated catheters into the individual ureters and then making

a radiograph. This was done in three of our series of duplication. It will be found more difficult and often impossible, however, to so diagnose the divisions of the ureter. With division of a single ureter above the meatus it will be found necessary to inject fluids opaque to the X-ray in order to demonstrate the condition. This we were able to do in two instances. Furthermore, the injected radiograph will usually aid us in ascertaining the existence of any complicating lesion such as stricture of the ureter and hydronephrosis.

The following complications were found in the cases of division of the ureter: hydronephrosis in a kidney with one ureter uniting with the ureter from the opposite kidney, 1; stone in ureter above point of lower division, 1; stricture in ureter at bladder, 1.

REFERENCES.

[1] Braasch: ANNALS OF SURG., Nov., 1910.
[2] Rovsing: Ztschr. f. Urologie, Bd. v, Heft 8, 1911.
[3] Anders: Am. Jour. Med. Sci., Mar., 1910.
[4] Monro: Bos. M. and S. Jour., Mar. 31, 1910.