Emmett's

CLINICAL UROGRAPHY

An Atlas and Textbook of Roentgenologic Diagnosis

WITTEN · MYERS · UTZ

VOLUME 2

Fourth Edition

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VOLUME II

Chapter 9

ANOMALIES OF THE GENITOURINARY TRACT

Campbell (1963) has stated that "anomalous development attains its highest incidence in the urogenital tract . . ." It is estimated that 35 per cent to 40 per cent of all maldevelopments are urogenital and that approximately 10 per cent of all human beings are born with some anomaly of the urogenital tract. Dees found that, in 9.6 per cent of 1,410 cases investigated urologically, urography disclosed some anomaly of the upper part of the urinary tract. This situation is not surprising when one considers the complicated embryologic development of the urinary tract. In the embryologic evolution of the kidney, *three* essential renal organs—the pronephros, the mesonephros, and the metanephros—develop in rapid succession. The pronephros and metanephros degenerate and become important parts of another system, the genital tract. Furthermore, the formation of the definitive kidney requires a delicate union of endodermal and mesodermal structures. Finally, the great number of changes that the fetal kidney normally undergoes both as to form and position are factors responsible for congenital anomalies. Although not every anomaly or congenital malformation constitutes a pathologic lesion or entity, it may be assumed from the clinical data at hand that practically every congenital malformation of the urinary tract is potentially a clinicopathologic entity.

It is apparent that this classification

CLINICAL CLASSIFICATION OF ANOMALIES

(This classification of possible anomalous states of the genitourinary system is by no means complete or all-inclusive and is confined to the anomalous conditions that can be demonstrated urographically.)

I. THE KIDNEY
- A. Anomalies in number
  - 1. Supernumerary kidney
  - 2. Agenesis and dysgenesis (unilateral and bilateral)
- B. Anomalies in size
  - 1. The small kidney (hypoplasia; atrophy)*
  - 2. Compensatory hypertrophy
- C. Anomalies in position
  - 1. Malrotation
  - 2. Ectopia and dystopia
- D. Anomalies in form
  - 1. Fusion
    - a. Crossed renal ectopia with fusion; horseshoe kidneys
- E. Anomalies in structure
  - 1. Polycystic disease†
  - 2. Multicystic disease†

*Also considered in Chapter 10.

†Considered in Chapter 13.

II. THE RENAL PELVIS AND URETER
- A. Anomalies in number
  1. Bifid, trifid, and multifid pelves
  2. Duplication of pelvis and ureter
- B. Anomalies of the ureteropelvic juncture‡
- C. Anomalies in position and form of ureter
  1. Rudimentary branched ureter
  2. Blind-ending duplication
  3. Ureteral diverticula
  4. Retrocaval ureter
  5. Congenital obstruction of ureter (stricture, valves, and aberrant blood vessels)
  6. Congenital "nonobstructive" ureterectasis (megaureter)‡
  7. Vesicoureteral reflux‡
- D. Anomalies of origin and termination of ureter
  1. Ectopic ureteral orifices
- E. Ureterocele; ectopic ureterocele

III. THE BLADDER AND URETHRA
- A. Anomalies of the urachus
- B. Anomalies of the bladder
  1. Exstrophy-epispadias complex
  2. Duplication of bladder
- C. Anomalies of the urethra
  1. Double urethra and accessory urethra
  2. Diverticulum of the male urethra

IV. DEEP PELVIC CYSTS
- A. Cysts of the müllerian duct
- B. Dilatation of prostatic utricle
- C. Cysts of the seminal vesicles
- D. Cysts of the prostate gland

V. CONGENITAL ABSENCE OF ABDOMINAL MUSCLES

VI. INTERSEXUALITY

VII. ECTOPIC ANUS (IMPERFORATE ANUS, ANAL ATRESIA, URETHRORECTAL "FISTULA")

‡Considered in Chapter 11.

could be enlarged and compounded if one were to list all the possible combinations of the many conditions enumerated. For instance, in the case of the ectopic ureteral orifice many combinations are possible that depend on whether the condition is unilateral or bilateral, is associated with duplication on one or both sides, involves one or more ureters, or is associated with ectopic or crossed ectopic kidneys. It is obviously impossible in a book of this kind to include urograms to demonstrate every type of anomaly of the kidney and ureter that has been described. Illustrations of the more common conditions will be shown which should be sufficient to permit recognition of almost any type of anomaly likely to be encountered.

Also, it should be readily apparent that in the subsequent discussion the outline cannot be followed rigidly because the various conditions often occur together with a myriad of possible combinations which at times may tax one's diagnostic acumen.

THE KIDNEY AND URETER

Embryology of the Kidney and Ureter*

THE KIDNEY: EARLY PHASES

The duct which drains the mesonephros (wolffian body) is called the *mesonephric* or *wolffian* duct. It originates as the pronephric duct which grows caudad, reaching and emptying into the cloaca at about the 4-mm stage. When mesonephric tubules become connected to the pronephric duct, its name is changed to mesonephric duct. The cloaca divides into a ventral segment, forming the bladder and

*Only the embryology pertinent to the anomalies discussed in this chapter is included here.

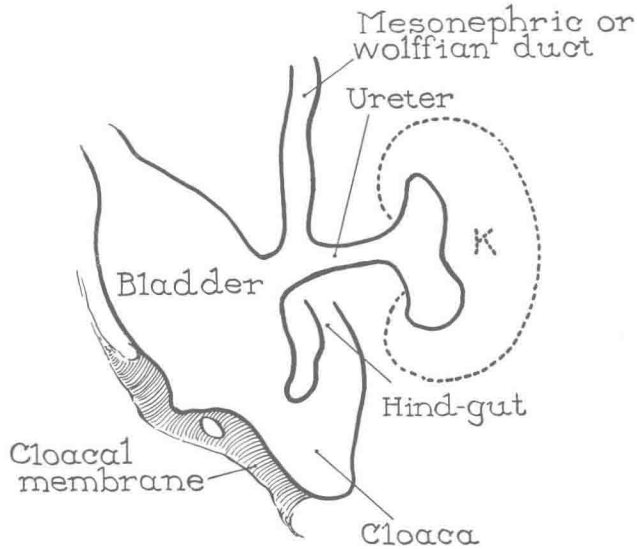


Figure 9–1. Sprouting of ureter from mesonephric or wolffian duct and its division into two branches which are forerunners of two major calyces. Renal mesenchyme, which caps branching ureter, has assumed definite "bean" form. (See text.) (Redrawn from Arey, L. B.)

the urogenital sinus, and a dorsal segment, which becomes the rectum. Even before the cloaca divides, a bud appears on the dorsal surface of the wolffian duct a short distance from the cloacal wall. This is called the *ureteral bud* and is the anlage of the adult ureter. The cranial end of the ureteral bud grows cephalad into a mass of undifferentiated mesoderm which soon becomes specialized to form the renal mesenchyme or anlage of the kidney proper. At approximately the 10-mm stage, the cephalic end of the ureteral bud divides into two branches which are the forerunners of the two major calyces (Fig. 9–1). At this stage the renal mesenchyme, which caps the branching ureter, has assumed a definite "bean" form. The two kidneys at this stage lie close together (almost in apposition and parallel to each other) and are at the level of the second sacral segment. Their upper poles reach to the brim of the true pelvis. The point of bifurcation of the ureteral bud becomes the future renal pelvis.

Further description of the subsequent branching to the ureter is best left to Arey, who said:

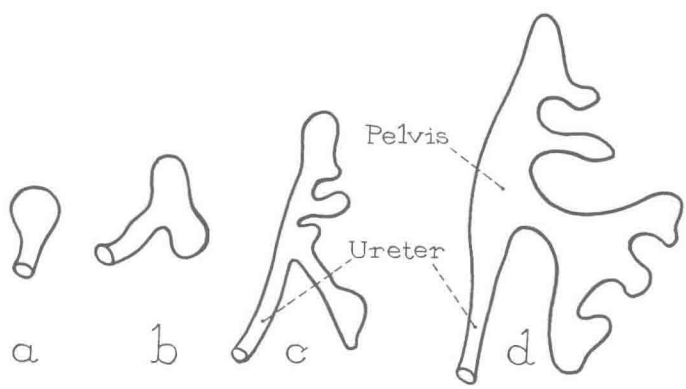


Figure 9–2. Branching of ureteral bud to form major and minor calyces. (See text.) (From Arey, L. B.)

Of the first two [primary divisions of the ureter] one is cranial, the other is caudal in position and between these, two others usually appear [Fig. 9–2]. From an ampullary enlargement at the end of each primary tubule sprout off two, three or four secondary tubules. These in turn give rise to tertiary tubules and the process is repeated until the fifth month of fetal life, when it is estimated that twelve generations of tubules have been developed. The pelvis and the primary and secondary tubules enlarge greatly during development. The two primary expansions become the *major calyces* and the secondary tubules opening into them form the *minor calyces*. The tubules of the third and fourth orders are taken up into the walls of the enlarged secondary tubules so that the tubules of the fifth order, twenty to thirty in number, open into the minor calyces as *papillary ducts*. The remaining orders of tubules constitute the *collecting tubules* which form the greater part of the medulla of the adult kidney.

The renal cortex is formed from the renal mesenchyme, which gives rise to the glomeruli and uriniferous tubules (proximal and distal convoluted tubules and loops of Henle).

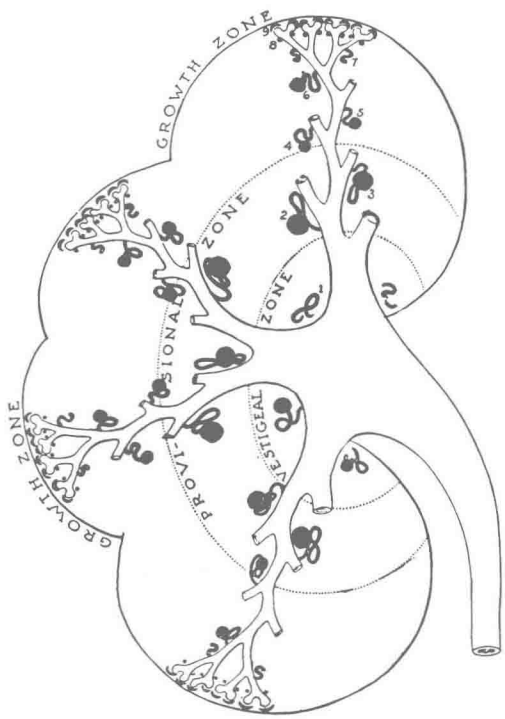


Figure 9–3. The three zones in development of uriniferous tubules. (From Kampmeier, O. F.)

Failure of the uriniferous tubules (originating from nephrogenic blastema) to connect with the collecting tubules (derived from the ureteric bud) is the most commonly accepted explanation of *polycystic disease of the kidneys*. However, Mc Kenna and Kampmeier, in 1934, advanced a different explanation based on Kampmeier's original concept of three zones in the developing cortex of the kidney, namely a vestigial zone, a provisional zone, and a growth zone (Fig. 9–3). Kampmeier expressed the belief that the tubules of the vestigial zone are rudimentary and disappear without a trace, those of the provisional zone unite with the collecting ducts only to break away from them again, and those of the growth zone form the definitive renal cortex. McKenna and Kampmeier stated that the uriniferous tubules of the provisional zone usually collapse and disappear after they lose their connections to the collecting tubules, but that some of them may persist and expand to form cysts. They explained all types of renal cysts (solitary, multiple, and polycystic disease) on this basis. [*This explanation of cyst formation has been challenged repeatedly in recent years, and it does not now appear to be correct. The subject is discussed in detail in chapter 13.*]

THE URETER

Returning now to consideration of the lower end of the ureter, it will be recalled that originally the ureter sprouted from the dorsal surface of the wolffian duct just proximal to its junction with the cloaca. The ureter next shifts its position to the lateral aspect of the wolffian duct. The lower end of the duct, from which the ureter arises, expands and through a rather complicated form of growth is absorbed as a part of the bladder in its vesicourethral portion so that the ureter and the mesonephric duct open separately (Fig. 9–4). This change is taking place at about the same time that the kid-

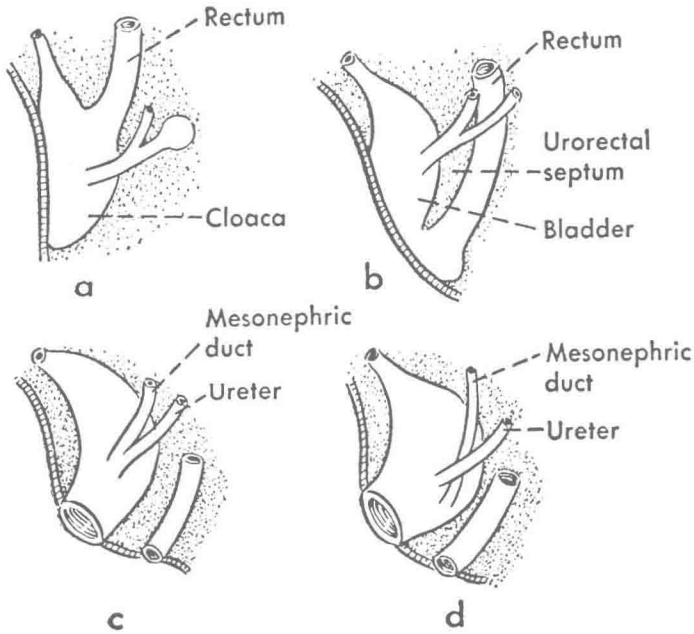


Figure 9–4. Method of subdivision of cloaca and separation of openings of mesonephric duct (vas deferens in male) and ureter. **A,** Primitive condition. **B,** Urorectal septum has separated rectum from urinary bladder. **C,** Common stem of mesonephric duct and ureter has been largely absorbed into wall of bladder. **D,** Differential growth of wall of bladder is carrying mesonephric duct distally toward urethra. (From Hollingshead, W. H.)

neys are undergoing rotation and ascent. The portion of the bladder that receives the ureters and wolffian ducts grows in an uneven manner so that the ureters migrate laterally and cranially while the distal ends of the wolffian ducts remain close together in the midline and appear to migrate somewhat distally, their orifices eventually being situated in the verumontanum in the distal portion of the floor of the prostatic urethra. Embryologically, therefore, an area bounded by the ureteral orifices and the mesonephric (ejaculatory) ducts is thought to be of mesodermal origin, whereas the remainder of the bladder is of entodermal origin. There is still some difference of opinion concerning this last statement, however. This area of mesonephric tissue in the male includes the trigone and proximal part of the prostatic urethra. In the female it includes the trigone and almost all of the urethra.

Anomalies in Number

"FREE" SUPERNUMERARY KIDNEY

One of the rarest anomalies encountered in the urinary tract is the free supernumerary kidney (Bacon; Carlson; Hanley; Kretschmer; Mills; Stewart; Swick) (Figs. 9–5 through 9–8). Up to five free supernumerary kidneys in one individual have been described. One must be exceedingly careful when making such a diagnosis not to confuse it with a duplicated pelvis or a complete duplication in a segment of a horseshoe kidney (see Figure 9–99).

Embryogenesis

The embryogenesis of the supernumerary kidney is similar to renal duplication. Two ureteric buds arise from different positions on the wolffian duct, reaching the

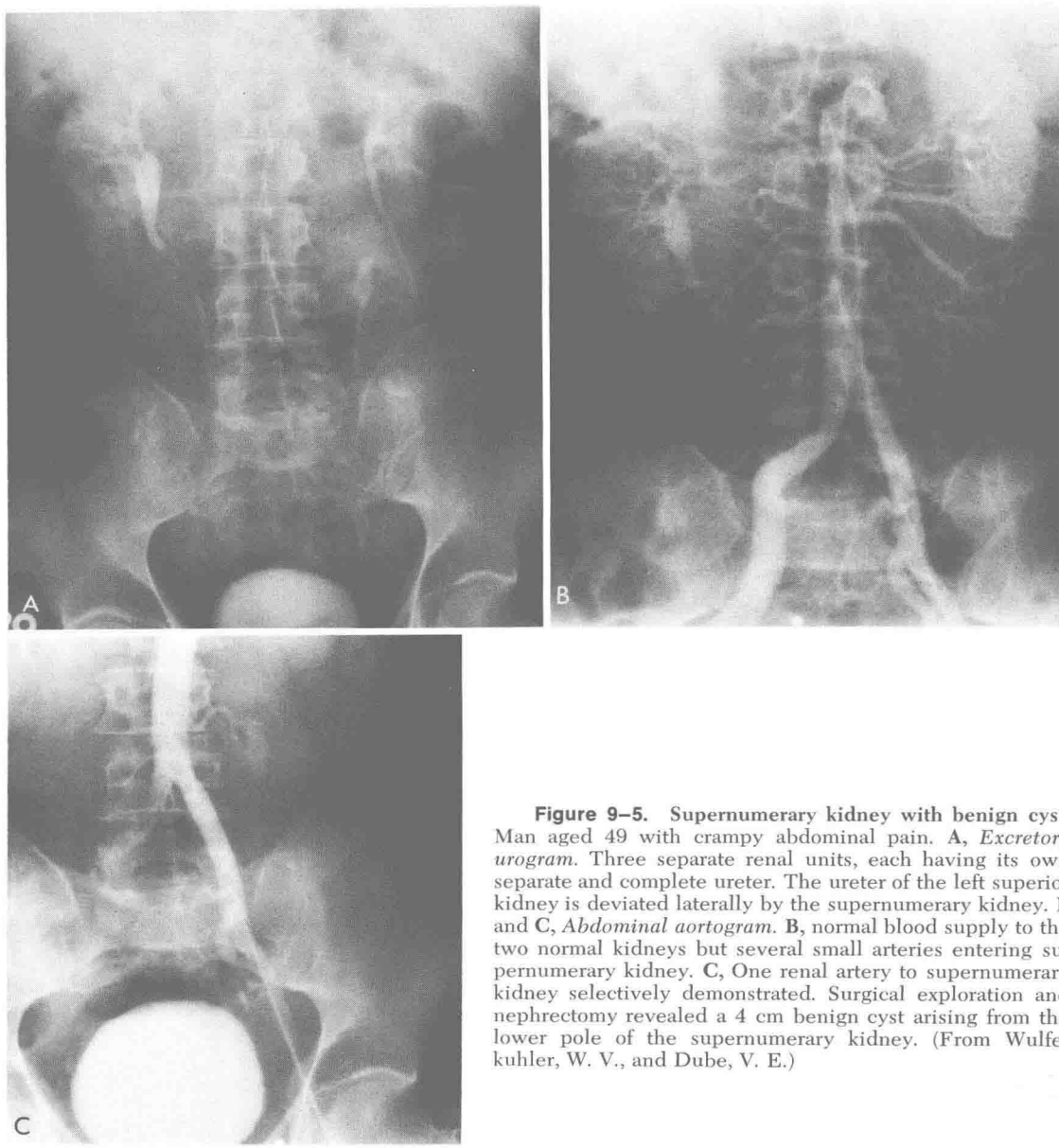


Figure 9–5. Supernumerary kidney with benign cyst. Man aged 49 with crampy abdominal pain. **A,** *Excretory urogram.* Three separate renal units, each having its own separate and complete ureter. The ureter of the left superior kidney is deviated laterally by the supernumerary kidney. **B** and **C,** *Abdominal aortogram.* **B,** normal blood supply to the two normal kidneys but several small arteries entering supernumerary kidney. **C,** One renal artery to supernumerary kidney selectively demonstrated. Surgical exploration and nephrectomy revealed a 4 cm benign cyst arising from the lower pole of the supernumerary kidney. (From Wulfekuhler, W. V., and Dube, V. E.)

nephrogenic mesenchyme (nephrogenic blastema) so divergent that two separate renal units develop. Usually the kidney in the inferior position is the supernumerary unit, but it may be superior. The supernumerary kidney is usually smaller and is often hypoplastic and histologically less organized than the normal kidney. For this reason there is often poor function, making the supernumerary kidney difficult to visualize by excretory urography. The ureter of the supernumerary kidney may join the normal ureter or enter the bladder separately. Rarely, the ureter of the upper kidney joins the lower kidney's pelvis (Fig. 9–9), or the ureter of the supernumerary kidney has an ectopic opening (Samuels and associates). Simultaneous anomalies of other parts of the urogenital tract as well as a high incidence of pathologic changes in

supernumerary kidneys have been reported (Wulfekuhler and Dube).

RENAL AGENESIS AND DYSGENESIS

Terminology

Ashley and Mostofi, in their comprehensive necropsy study on this subject, have suggested the following terminology: (1) If no vestige of renal tissue is formed, the condition is called *agenesis.* (2) If the kidney is represented by a nodule of tissue that bears no morphologic or histologic resemblance to normal renal parenchyma, it is called *dysgenesis.* (3) If the kidney is tiny but otherwise similar to a normal organ, it is called *hypoplasia.* Other terms that have been used in the literature for the latter two terms are *aplasia, dysplasia,* and *congenital atrophy.*

Embryogenesis

Renal agenesis and dysgenesis result from a failure of the ureteric bud to make contact with the nephrogenic blastema at the proper time. The reason for this failure may be one of several: (1) failure of the ureteric bud to form, (2) failure of the bud to reach the blastema before it migrates upward, (3) failure of the wolffian duct to

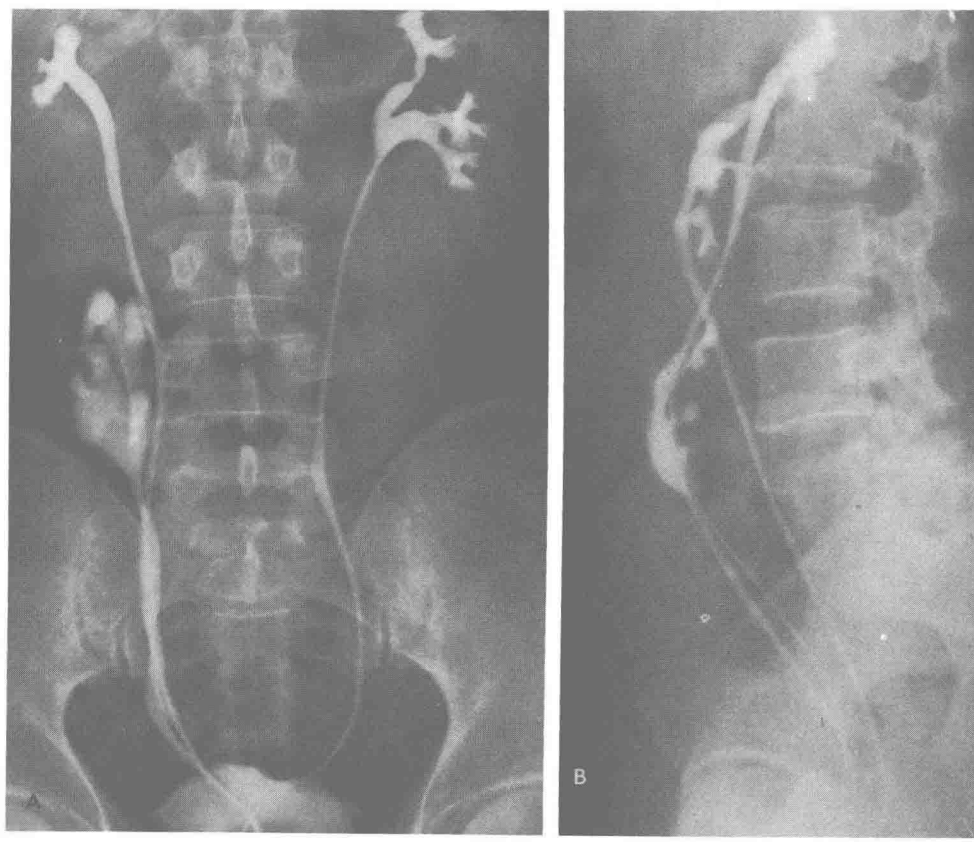
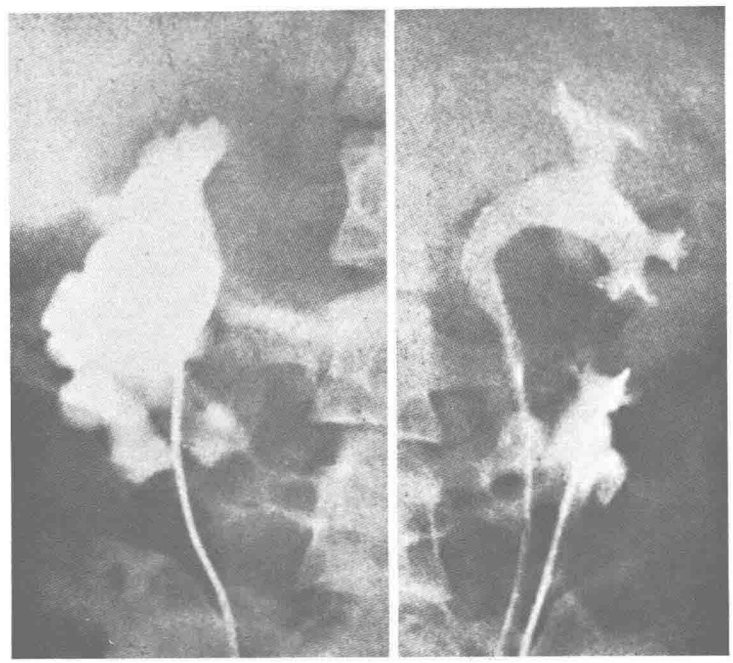


Figure 9–6. Supernumerary kidney associated with pyelectasis on right side, opposite fourth and fifth lumbar vertebrae. This is associated with **complete duplication of right ureter and two ureteral orifices in bladder.** Little if any function remains in right supernumerary kidney. **A,** *Bilateral retrograde pyelogram,* showing three separate kidneys. **B,** *Lateral film.* Exploration revealed supernumerary kidney 6 to 7 cm long on right side, lying in groove between psoas muscle and vertebral bodies in approximate area of right sacroiliac articulation. Upper right kidney and left kidney were palpated and found to be normal except that right kidney was about one third smaller than normal. (From Stewart, C. M.)



A B

Figure 9–7. Horseshoe kidney associated with supernumerary kidney. A, *Right retrograde pyelogram.* Larger half of horseshoe kidney is on right, smaller on left. **Moderate pyelocaliectasis on right. B,** *Left retrograde pyelogram.* Pelvis supplying left half of horseshoe is smaller than right. Above is supernumary kidney which at surgical exploration was freely movable in all directions and separate from horseshoe kidney; separate vascular pedicle could also be palpated. (From Hanley, H. G.)

develop, or (4) the absence of the metanephrogenic mesenchyme. When there is failure of the urogenital ridge to form, there is absence of all internal genital and upper urinary tract structures, including the trigone on the affected side. When the ureteric bud fails to reach or enter the nephrogenic blastema at the proper time, there is either absence or dysgenesis of the kidney. The ureter is absent in most cases of renal agenesis and is usually atretic or absent when dysgenesis is present.

Incidence

Autopsy Material. It is impossible to give any firm data concerning the incidence of renal agenesis. Autopsy studies provide the most significant data. Clinical data are notoriously inaccurate; however, since excretory urography has become so widely used, clinical data are becoming more significant. Of course, *bilateral agenesis* is extremely rare, incompatible with life, and, therefore, of little clinical importance. Potter described a characteristic facial appearance in infants with bilateral renal agenesis. The face shows an increased space between the eyes and a prominent fold arising at the inner canthus which sweeps downward and laterally below the eyes. In addition, there are a flattened nose, receding chin, and low-set ears. *Unilateral agenesis* appears to be not too uncommon. Most urologists encounter this condition frequently enough to constantly have it in mind.

From a collection of 245,000 autopsy protocols in the files of the Armed Forces Institute of Pathology, Ashley and Mostofi found 364 cases of renal agenesis or dysgenesis as follows: bilateral agenesis—47 cases; unilateral agenesis—240 cases (including 8 cases of crossed ectopia);

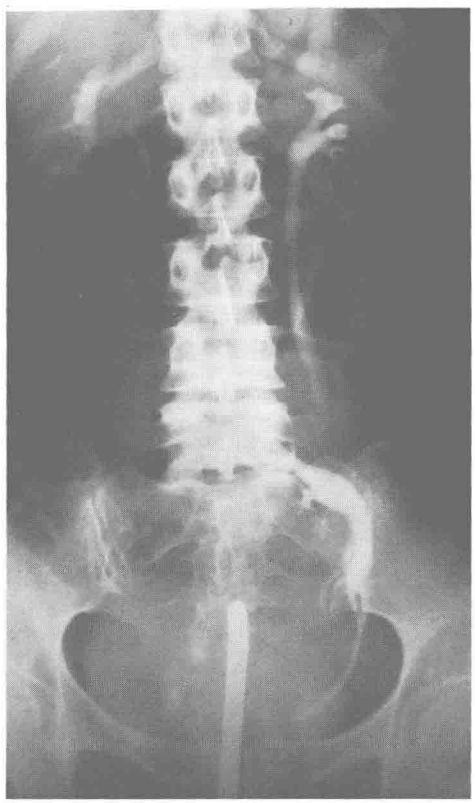


Figure 9–8. *Retrograde pyelogram.* **Supernumerary left kidney** overlies left sacro-iliac synchondrosis. Diagnosis was confirmed at laparotomy. (Courtesy of Dr. W. E. Kittredge.)

bilateral dysgenesis—11 cases; unilateral dysgenesis—57 cases; and mixed anomaly (dysgenesis on one side and agenesis on the other)—9 cases.

Because of the military nature of the population from which the study was drawn (high proportion of young men), data concerning sex incidence and age are not significant.

In 19,046 autopsies of children, Campbell (1963) found bilateral agenesis in seven cases (ratio of 1:2,721); whereas, in another series of 51,880 autopsies, there were 94 cases of unilateral agenesis (ratio of 1:552).

Clinical Material. Collins collected 581 cases of unilateral agenesis from the world literature. Of the 581 patients, 281 (48.3 per cent) were male and 231 (39.7 per cent) were female; the sex was not given in 69 cases.

Braasch and Merricks were able to find 69 cases of unilateral agenesis in the records of the Mayo Clinic from 1909 through 1937. Only 27 of these were verified by operation or autopsy. Longo and Thompson in a later clinical study found an additional 94 cases seen at the Mayo Clinic in a 12-year period, 1938 through 1949. Only five of these cases were verified by operation, but all were exhaustively and carefully studied. The authors stated that unilateral renal agenesis is found in about 1 of every 1,000 autopsies and clinically in about 1 of every 500 urologic patients.

In the past, unilateral agenesis has been considered to occur more commonly in females than in males, but in Longo and Thompson's series 60.6 per cent of the cases involved males and only 39.3 per cent females. It was their opinion that because females have a greater frequency of complications and associated anomalies (especially genital) than males, they are led to seek medical consultation more often.

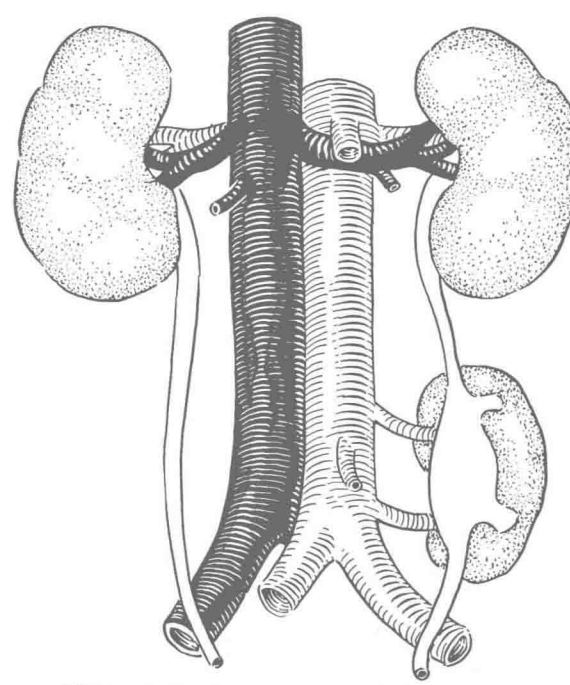


Figure 9–9. Supernumerary kidney. (Redrawn from Kretschmer, H. L. By permission of Surg., Gynec. & Obst.)