congenital and acquired atrophic kidneys may be impossible, but a truly hypoplastic kidney shows no scars and has a reduced number of renal lobes and pyramids, usually six or fewer.

Ectopic Kidneys. The development of the metanephros into the kidneys may occur in ectopic foci. These kidneys lie either just above the pelvic brim or sometimes within the pelvis. They are usually normal or slightly small in size but otherwise are not remarkable. Because of their abnormal position, kinking or tortuosity of the ureters may cause obstruction to urinary flow, which predisposes to bacterial infections.

Horseshoe Kidneys. Fusion of the upper (10%) or lower poles (90%) of the kidneys produces a horseshoe-shaped structure that is continuous across the midline anterior to the great vessels. This anomaly is found in 1 in 500 to 1000 autopsies.

Cystic Diseases of the Kidney

Cystic diseases of the kidney are heterogeneous, comprising hereditary, developmental, and acquired disorders. They are important for several reasons: (1) They are reasonably common and often represent diagnostic problems for clinicians, radiologists, and pathologists; (2) some forms, such as adult polycystic kidney disease, are major causes of chronic kidney disease; and (3) they can occasionally be confused with malignant tumors. A useful classification of renal cysts is as follows:

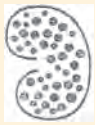
- Polycystic kidney disease
  - Autosomal dominant (adult) polycystic disease
  - Autosomal recessive (childhood) polycystic disease
- Medullary cystic disease
  - Medullary sponge kidney
  - Nephronophthisis
- Multicystic renal dysplasia
- Acquired (dialysis-associated) cystic disease
- Localized (simple) renal cysts
- Renal cysts in hereditary malformation syndromes (e.g., tuberous sclerosis)
- Glomerulocystic disease
- Extraparenchymal renal cysts (pyelocalyceal cysts, hilar lymphangitic cysts)

Only the more important of the cystic diseases are discussed later. Table 20-11 summarizes the characteristic features of the principal renal cystic diseases.

Autosomal Dominant (Adult) Polycystic Kidney Disease

Autosomal dominant (adult) polycystic kidney disease is a hereditary disorder characterized by multiple expanding cysts of both kidneys that ultimately destroy the renal parenchyma and cause renal failure. It is a common condition affecting roughly 1 of every 400 to 1000 live births and accounting for about 5% to 10% of cases of end-stage renal disease requiring transplantation or dialysis. The inheritance pattern is autosomal dominant with high

Table 20-11 Summary of Renal Cystic Diseases

Disease	Inheritance	Pathologic Features	Clinical Features or Complications	Typical Outcome	Diagrammatic Representation
Adult polycystic kidney disease	Autosomal dominant	Large multicystic kidneys, liver cysts, berry aneurysms	Hematuria, flank pain, urinary tract infection, renal stones, hypertension	Chronic renal failure beginning at age 40-60 years	
Childhood polycystic kidney disease	Autosomal recessive	Enlarged, cystic kidneys at birth	Hepatic fibrosis	Variable, death in infancy or childhood	
Medullary sponge kidney	None	Medullary cysts on excretory urography	Hematuria, urinary tract infection, recurrent renal stones	Benign	
Familial juvenile nephronophthisis	Autosomal recessive	Corticomedullary cysts, shrunken kidneys	Salt wasting, polyuria, growth retardation, anemia	Progressive renal failure beginning in childhood	
Adult-onset medullary cystic disease	Autosomal dominant	Corticomedullary cysts, shrunken kidneys	Salt wasting, polyuria	Chronic renal failure beginning in adulthood	
Multicystic renal dysplasia	None	Irregular kidneys with cysts of variable size	Association with other renal anomalies	Renal failure if bilateral, surgically curable if unilateral	
Acquired renal cystic disease	None	Cystic degeneration in end-stage kidney disease	Hemorrhage, erythrocytosis, neoplasia	Dependence on dialysis	
Simple cysts	None	Single or multiple cysts in normal-sized kidneys	Microscopic hematuria	Benign