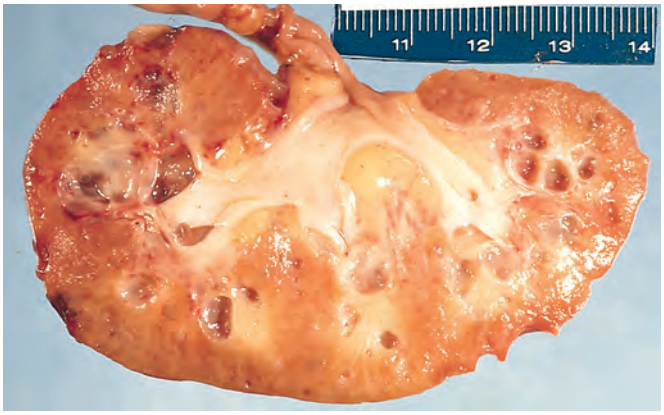
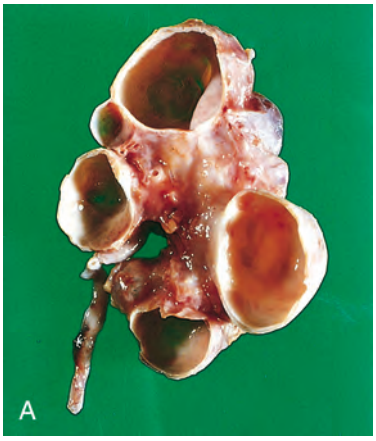


Figure 20-45 Medullary cystic disease. Cut section of kidney showing cysts at the corticomedullary junction and in the medulla.

renal failure, a positive family history, and chronic tubulointerstitial nephritis on biopsy.

Multicystic Renal Dysplasia

Dysplasia is a sporadic disorder that can be unilateral or bilateral and is almost always cystic. The kidney is usually enlarged, extremely irregular, and multicystic (Fig. 20-46*A*).



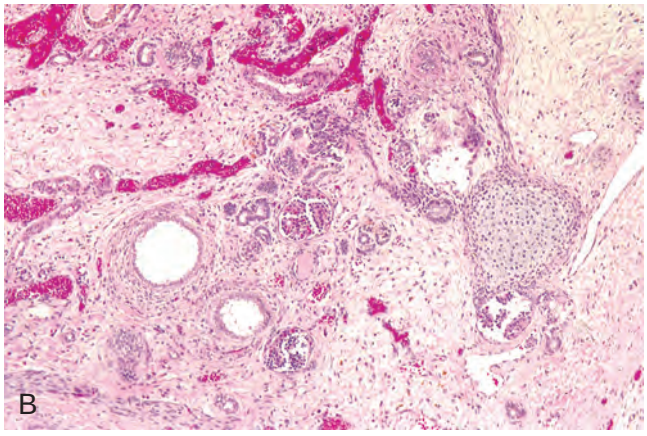


Figure 20-46 Multicystic renal dysplasia. **A,** Gross appearance. **B,** Histologic section showing disorganized architecture, dilated tubules with cuffs of primitive stroma, and an island of cartilage (Hematoxylin and eosin stain). (**A,** Courtesy Dr. D. Schofield, Children's Hospital, Los Angeles, Calif.; **B,** courtesy Dr. Laura Finn, Children's Hospital, Seattle, Wash.)

The cysts vary in size from several millimeters to centimeters in diameter. On histologic examination, they are lined by flattened epithelium. Although normal nephrons are present, many have immature collecting ducts. The characteristic histologic feature is the presence of islands of undifferentiated mesenchyme, often with cartilage, and immature collecting ducts (Fig. 20-46*B*). Most cases are associated with ureteropelvic obstruction, ureteral agenesis or atresia, and other anomalies of the lower urinary tract.

When unilateral, the dysplasia may mimic a neoplasm and lead to surgical exploration and nephrectomy. The opposite kidney functions normally, and such patients have an excellent prognosis after surgical removal of the affected kidney. In bilateral multicystic renal dysplasia, renal failure may ultimately result.

Acquired (Dialysis-Associated) Cystic Disease

Patients with end-stage renal disease who have undergone prolonged dialysis sometimes show numerous cortical and medullary renal cysts. The cysts measure 0.1 to 4 cm in diameter, contain clear fluid, are lined by either hyperplastic or flattened tubular epithelium, and often contain calcium oxalate crystals. They probably form as a result of obstruction of tubules by interstitial fibrosis or by oxalate crystals. Most are asymptomatic, but sometimes the cysts bleed, causing hematuria. There is a 12- to 18-fold increased risk of renal cell carcinoma, which develops in 7% of dialyzed patients observed for 10 years.

Simple Cysts

Simple cysts may be single or multiple and usually involve the cortex. They are commonly 1 to 5 cm but may reach 10 cm or more in size. They are translucent, lined by a gray, glistening, smooth membrane, and filled with clear fluid. On microscopic examination these membranes are composed of a single layer of cuboidal or flattened cuboidal epithelium, which in many instances may be completely atrophic.

Simple cysts are common postmortem findings without clinical significance. On occasion, hemorrhage into them may cause sudden distention and pain, and calcification of the hemorrhage may give rise to bizarre radiographic shadows. The main importance of cysts lies in their differentiation from kidney tumors. Radiologic studies show that in contrast to renal tumors, renal cysts have smooth contours, are almost always avascular, and give fluid rather than solid signals on ultrasonography.

KEY CONCEPTS

Cystic Diseases

- Autosomal dominant polycystic kidney disease accounts for a small yet significant subset of end stage renal disease.
- Ciliopathies or abnormalities of the cilium-centrosome complex underlie the major cystic kidney diseases, including polycystic kidney disease (both autosomal dominant and autosomal recessive forms), medullary cystic kidney disease, and nephronophthisis.