Endemic Diseases

Initially thought to be restricted to Scandinavian countries, and thus termed Scandinavian acute hemorrhagic interstitial nephritis, Nephropathia epidemica has been shown to have a more universal occurrence. It therefore has been more appropriately renamed hemorrhagic fever with renal syndrome. As a rule the disease presents as a reversible acute tubulointerstitial nephritis but can progress to a chronic form. It is caused by a rodent-transmitted virus of the Hantavirus genus of the Bunyaviridae family, the so-called Hantaan virus. Humans appear to be infected by respiratory aerosols contaminated by rodent excreta. Antibodies to the virus are detected in the serum, and viruslike structures have been demonstrated in the kidneys of persons infected with the virus.

Tubulointerstitial nephropathy caused by viral infection also has been reported in polyomavirus, cytomegalovirus, herpes simplex virus, human immunodeficiency virus, infectious mononucleosis, and Epstein-Barr virus.

Hereditary Diseases

Hereditary Nephritis

Hereditary nephritis or Alport’s syndrome, in which a mutation in the encoding gene localized to the X chromosome results in a defect in the \( \alpha-5 \) chain of type IV collagen.
Papillary Necrosis

A, Renal papillary necrosis. The arrow points to the region of a sloughed necrotic papilla. B, Whole mount of a necrotic papilla. Arrows delineate focal necrosis principally affecting the medullary inner stripe. Renal papillary necrosis (RPN) develops in a variety of diseases that cause chronic tubulointerstitial nephropathy in which the lesion is more severe in the inner medulla. The basic lesion affects the vasculature with consequent focal or diffuse ischemic necrosis of the distal segments of one or more renal pyramids. In the affected papilla, the sharp demarcation of the lesion and coagulative necrosis seen in the early stages of the disease closely resemble those of infarction. The fact that the necrosis is anatomically limited to the papillary tips can be attributed to a variety of features unique to this site, especially those affecting the vasculature. The renal papilla receives its blood supply from the vasa recta. Measurements of medullary blood flow notwithstanding, it should be noted that much of the blood flow in the vasa recta serves the countercurrent exchange mechanism. Nutrient blood supply is provided by small capillary vessels that originate in each given region. The net effect is that the blood supply to the papillary tip is less than that to the rest of the medulla, hence its predisposition to ischemic necrosis.

The necrotic lesions may be limited to only a few of the papillae or may involve several of the papillae in either one or both kidneys. The lesions are bilateral in most patients. In patients with involvement of one kidney at the time of initial presentation, RPN will develop in the other kidney within 4 years, which is not unexpected because of the systemic nature of the diseases associated with RPN. RPN may be unilateral in patients in whom predisposing factors (such as infection and obstruction) are limited to one kidney.

Azotemia may be absent even in bilateral papillary necrosis, because it is the total number of papillae involved that ultimately determines the level of renal insufficiency that develops. Each human kidney has an average of eight pyramids, such that even with bilateral RPN affecting one papilla or two papillae in each kidney, sufficient unaffected renal lobules remain to maintain an adequate level of renal function.

As a rule, RPN is a disease of an older age group, the average age of patients being 53 years. Nearly half of cases occur in persons over 60 years of age. More than 90% of cases occur in persons over 40 years of age, except for those caused by sickle cell hemoglobinopathy. RPN is much less common in children, in whom the chronic conditions associated with papillary necrosis are rare. However, RPN does occur in children in association with hypoxia, dehydration, and septicemia.