The kidneys are affected by a variety of systemic vasculitides [1,2]. This is not surprising given the numerous and varied types of vessels in the kidneys. The clinical manifestations and even the pathologic expressions of vasculitis often are not specific for a particular diagnostic category of vasculitis. An accurate precise diagnosis usually requires the integration of many different types of data, including clinical signs and symptoms, associated diseases (eg, asthma, systemic lupus erythematosus, rheumatoid arthritis, hepatitis virus, polymyalgia rheumatica), vascular distribution (ie, types and locations of involved vessels), histologic pattern of inflammation (eg, granulomatous versus necrotizing), immunopathologic features (eg, presence and composition of vascular immunoglobulin deposits), and serologic findings (eg, cryoglobulins, hypocomplementemia, hepatitis B antibodies, hepatitis C antibodies, antineutrophil cytoplasmic autoantibodies, anti-glomerular basement membrane [GBM] antibodies, antinuclear antibodies). Specific diagnosis of a vasculitis is very important because the prognosis and appropriate therapy vary substantially among different types of vasculitis.

A general overview of the major categories of vasculitis that affect the kidneys is presented. The focus is primarily on polyarteritis nodosa, Henoch-Schönlein purpura, Wegener’s granulomatosis, and microscopic polyangiitis.
# 2.2 Systemic Diseases and the Kidney

## Overview

### SELECTED CATEGORIES OF VASCULITIS

- Large vessel vasculitis
  - Giant cell arteritis
  - Takayasu arteritis
- Medium-sized vessel vasculitis
  - Polyarteritis nodosa
  - Kawasaki disease
- Small vessel vasculitis
  - ANCA small vessel vasculitis
  - Microscopic polyangiitis
  - Wegener’s granulomatosis
  - Churg-Strauss syndrome
  - Immune complex small vessel vasculitis
    - Henoch-Schönlein purpura
    - Cryoglobulinemic vasculitis
    - Lupus vasculitis
    - Serum sickness vasculitis
    - Infection-induced immune complex vasculitis
  - Anti–GBM small vessel vasculitis
  - Goodpasture’s syndrome

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**FIGURE 2-1**

Many different approaches to categorizing vasculitis exist. We use the approach adopted by the Chapel Hill International Consensus Conference on the Nomenclature of Systemic Vasculitis [3]. The Chapel Hill System divides vasculitides into those that have a predilection for large arteries (i.e., the aorta and its major branches), medium-sized vessels (i.e., main visceral arteries), and small vessels (predominantly capillaries, venules, and arterioles, and occasionally, small arteries). However, there is so much overlap in the size of the vessel involved by different vasculitides that other criteria are very important for precise diagnosis, especially when distinguishing among the different types of small vessel vasculitis. ANCA—antineutrophil cytoplasmic antibody.

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**FIGURE 2-2**

Predominant distributions of renal vascular involvement. This diagram depicts the predominant distributions of renal vascular involvement by large, medium-sized, and small vessel vasculitides [2]. Note that all three categories may affect arteries, although arteries are least often affected by the small vessel vasculitides and often are not involved at all by this category of vasculitis. By the Chapel Hill definitions, glomerular involvement (i.e., glomerulonephritis) is confined to the small vessel vasculitides, which provides a concrete criterion for separating the diseases in this category from those in the other two categories [3].
Vasculitis (Polyarteritis Nodosa, Microscopic Polyangiitis, Wegener’s Granulomatosis, Henoch-Schönlein Purpura)

**RENAL INJURY CAUSED BY DIFFERENT CATEGORIES OF VASCULITIS**

Large vessel vasculitis  
Ischemia causing renovascular hypertension (uncommon)

Medium-sized vessel vasculitis  
Renal infarcts (frequent)  
Hemorrhage (uncommon)  
and rupture (rare)

ANCA small vessel vasculitis  
Pauci-immune crescentic glomerulonephritis (common)  
Arcuate and interlobular arteritis (occasional)  
Medullary angiitis (uncommon)  
Interstitial granulomatous inflammation (rare)

Immune complex small vessel vasculitis  
Immune complex proliferative or membranoproliferative glomerulonephritis with or without crescents (common)  
Arteriolitis and interlobular arteritis (rare)

Anti-GBM small vessel vasculitis  
Crescentic glomerulonephritis (common)  
Extraglomerular vasculitis (only with concurrent ANCA disease)

**FIGURE 2-3**

The type of renal vessel involved by a vasculitis determines the resultant renal dysfunction. Large vessel vasculitides cause renal dysfunction by injuring the renal arteries and the aorta adjacent to the renal artery ostia. These injuries result in reduced renal blood flow and resultant renovascular hypertension. Medium-sized vessel vasculitides most often affects lobar, arcuate, and interlobular arteries, resulting in infarction and hemorrhage. Small vessel vasculitides most often affect the glomerular capillaries (ie, cause glomerulonephritis), but some types (especially the antineutrophil cytoplasmic antibody vasculitides) may also affect extraglomerular parenchymal arterioles, venules, and capillaries. Anti-GBM disease is a form of vasculitis that involves only capillaries in glomeruli or pulmonary alveoli, or both. This category of vasculitis is considered in detail separately in this Atlas.

**FIGURE 2-4**

The two major categories of large vessel vasculitis, giant cell (temporal) arteritis and Takayasu arteritis, are both characterized pathologically by granulomatous inflammation of the aorta, its major branches, or both. The most reliable criterion for distinguishing between these two disease is the younger age of patients with Takayasu arteritis compared with giant cell arteritis [3]. The presence of polymyalgia rheumatica supports a diagnosis of giant cell arteritis. Clinically significant renal disease is more commonly associated with Takayasu arteritis than giant cell arteritis, although pathologic involvement of the kidneys is a frequent finding with both conditions [4,5].