The primary glomerulopathies are those disorders that affect glomerular structure, function, or both in the absence of a multisystem disorder. The clinical manifestations are predominantly the consequence of the glomerular lesion (such as proteinuria, hematuria, and reduced glomerular filtration rate). The combination of clinical manifestations leads to a variety of clinical syndromes. These syndromes include acute glomerulonephritis; rapidly progressive glomerulonephritis; chronic renal failure; the nephrotic syndrome or “asymptomatic” hematuria, proteinuria, or both.
2.2 Glomerulonephritis and Vasculitis

**CLINICAL SYNDROMES OF GLOMERULAR DISEASE**

- Acute glomerulonephritis
- Rapidly progressive glomerulonephritis
- Chronic glomerulonephritis
- Nephrotic syndrome
- "Asymptomatic" hematuria, proteinuria, or both

**PRIMARY GLOMERULAR LESIONS**

- Minimal change disease
- Focal segmental glomerulosclerosis with hyalinosis
- Membranous glomerulonephritis
- Membranoproliferative glomerulonephritis
- Mesangial proliferative glomerulonephritis
- Crescentic glomerulonephritis
- Immunoglobulin A nephropathy
- Fibrillary and immunotactoid glomerulonephritis
- Collagenofibrotic glomerulopathy
- Lipoprotein glomerulopathy

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**FIGURE 2-1**

Each of these syndromes arises as a consequence of disturbances of glomerular structure and function. Acute glomerulonephritis consists of the abrupt onset of hematuria, proteinuria, edema, and hypertension. Rapidly progressive glomerulonephritis is characterized by features of nephritis and progressive renal insufficiency. Chronic glomerulonephritis features proteinuria and hematuria with indolent progressive renal failure. Nephrotic syndrome consists of massive proteinuria (>3.5 g/d in adults), hypoalbuminemia with edema, lipiduria, and hyperlipidemia. "Asymptomatic" hematuria, proteinuria, or both is not associated initially with renal failure or edema. Each of these syndromes may be complicated by hypertension.

**FIGURE 2-2**

Age-associated prevalence of various glomerular lesions in nephrotic syndrome. This schematic illustrates the age-associated prevalence of various diseases and glomerular lesions among children and adults undergoing renal biopsy for evaluation of nephrotic syndrome (Guy's Hospital and the International Study of Kidney Disease in Children) [1]. Both the systemic and primary causes of nephrotic syndrome are included. (Diabetes mellitus with nephropathy is underrepresented because renal biopsy is seldom needed for diagnosis.) The bar on the left summarizes the prevalence of various lesions in children aged 0 to 16 years; the bar on the right summarizes the prevalence of various lesions in adults aged 16 to 80 years. Note the high prevalence of minimal change disease in children and the increasing prevalence of membranous glomerulonephritis in the age group of 16 to 60 years. FSGS—focal segmental glomerulosclerosis; MCGN—mesangiocapillary glomerulonephritis. (From Cameron [1]; with permission.)

**FIGURE 2-3**

The primary glomerular lesions.
Minimal Change Disease

FIGURE 2-4
Light and electron microscopy in minimal change disease (lipoid nephrosis). A. This glomerulopathy, one of many associated with nephrotic syndrome, has a normal appearance on light microscopy. No evidence of antibody (immune) deposits is seen on immunofluorescence. B. Effacement (loss) of foot processes of visceral epithelial cells is observed on electron microscopy. This last feature is the major morphologic lesion indicative of massive proteinuria.

Minimal change disease is considered to be the result of glomerular capillary wall damage by lymphokines produced by abnormal T cells. This glomerulopathy is the most common cause of nephrotic syndrome in children (>70%) and also accounts for approximately 20% of adult patients with nephrotic syndrome. This glomerulopathy typically is a corticosteroid-responsive lesion, and usually has a benign outcome with respect to renal failure.

FIGURE 2-5
Therapeutic response in minimal change disease. This graph illustrates the cumulative complete response rate (absence of abnormal proteinuria) in patients of varying ages in relation to type and duration of therapy [1]. Note that most children with minimal change disease respond to treatment within 8 weeks. Adults require prolonged therapy to reach equivalent response rates. Number of patients are indicated in parentheses. (From Cameron [2]; with permission.)

FIGURE 2-6
Cyclophosphamide in minimal change disease. One of several controlled trials of cyclophosphamide therapy in pediatric patients that pursued a relapsing steroid-dependent course is illustrated. Note the relative freedom from relapse when children were given a 12-week course of oral cyclophosphamide. An 8-week course of chlorambucil (0.15–0.2 mg/kg/d) may be equally effective. (From Arbeitsgemeinschaft für pediatrische nephrologie [3]; with permission.)