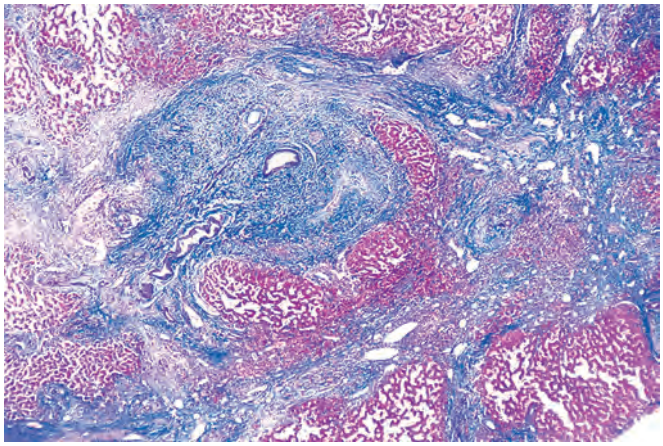

Figure 8-36 Trichrome stain of liver shows a gumma (scar), stained blue, caused by tertiary syphilis (the hepatic lesion is also known as hepar lobatum).

Syphilitic gummas are white-gray and rubbery, occur singly or multiply, and vary in size from microscopic lesions resembling tubercles to large tumor-like masses. They occur in most organs but particularly in skin, subcutaneous tissue, bone, and joints. In the liver, scarring as a result of gummas may cause a distinctive hepatic lesion known as hepar lobatum (Fig. 8-36). On histologic examination, the gummas have centers of coagulated, necrotic material and margins composed of plump, palisading macrophages and fibroblasts surrounded by large numbers of mononuclear leukocytes, chiefly plasma cells. Treponemes are scant in gummas and are difficult to demonstrate.

The rash of **congenital syphilis** is more severe than that of adult secondary syphilis. It is a bullous eruption of the palms and soles of the feet associated with epidermal sloughing. **Syphilitic osteochondritis and periostitis** affect all bones, but lesions of the nose and lower legs are most distinctive. Destruction of the vomer causes collapse of the bridge of the nose and, later on, the characteristic saddle nose deformity. Periostitis of the tibia leads to excessive new bone growth on the anterior surfaces and anterior bowing, or saber shin. There is also widespread disturbance in endochondral bone formation. The epiphyses become widened as the cartilage overgrows, and cartilage is found in displaced islands within the metaphysis.

The **liver** is often severely affected in congenital syphilis. Diffuse fibrosis permeates lobules to isolate hepatic cells into small nests, accompanied by the characteristic lymphoplasmacytic infiltrate and vascular changes. Gummas are occasionally found in the liver, even in early cases. The **lungs** may be affected by a diffuse interstitial fibrosis. In the syphilitic stillborn, the lungs appear pale and airless (pneumonia alba). The generalized spirochetemia may lead to diffuse interstitial inflammatory reactions in virtually any other organ (e.g., the pancreas, kidneys, heart, spleen, thymus, endocrine organs, and CNS).

The late manifestations of congenital syphilis include a distinctive **triad of interstitial keratitis, Hutchinson teeth, and eighth-nerve deafness**. In addition to interstitial keratitis, the ocular changes include choroiditis and abnormal retinal pigmentation. Hutchinson teeth are small incisors shaped like a screwdriver or a peg, often with notches in the enamel. Eighth-nerve deafness and optic nerve atrophy develop secondary to meningovascular syphilis.

Lyme Disease

Lyme disease is a common arthropod-borne illness caused by the spirochete, *Borrelia burgdorferi*, which can be localized or disseminated with a tendency to cause persistent chronic arthritis. It is named for the Connecticut town where there was an epidemic of arthritis associated with skin erythema in the mid-1970s. It is caused by several subspecies of the spirochete *Borrelia burgdorferi,* which is transmitted from rodents to people by *Ixodes* deer ticks. Lyme disease is endemic in the United States, Europe, and Japan. In the United States there were approximately 33,000 confirmed and probable cases reported in 2011. Most cases occur in the northeastern states and the upper Midwest. In endemic areas, *B. burgdorferi* infects up to 50% of ticks, which may also be infected with *Ehrlichia* and *Babesia*. Serology is the main method of diagnosis, but PCR can be done on infected tissue.

Lyme disease involves multiple organ systems and is divided into three stages (Fig. 8-37).

- In *stage 1 (localized infection)* spirochetes multiply and spread in the dermis at the site of a tick bite, causing an expanding area of redness, often with a pale center. This lesion, called *erythema migrans*, may be accompanied by fever and lymphadenopathy. The rash spontaneously disappears in 4 to 12 weeks.
- In *stage 2 (disseminated infection)* spirochetes spread hematogenously throughout the body and cause secondary skin lesions, lymphadenopathy, migratory joint and muscle pain, cardiac arrhythmias, and meningitis often associated with cranial nerve involvement.
- *Stage 3 (persistent infection)* manifests many months after the tick bite. *B. burgdorferi* usually causes a chronic arthritis sometimes with severe damage to large joints. Less often, patients will have polyneuropathy and encephalitis that vary from mild to debilitating.

Pathogenesis. *B. burgdorferi* does not produce LPS or exotoxins that damage the host. **Much of the pathology associated with *B. burgdorferi* is thought to be secondary to the immune response against the bacteria and the inflammation that accompanies it.** The initial immune response is stimulated by binding of bacterial lipoproteins to TLR2

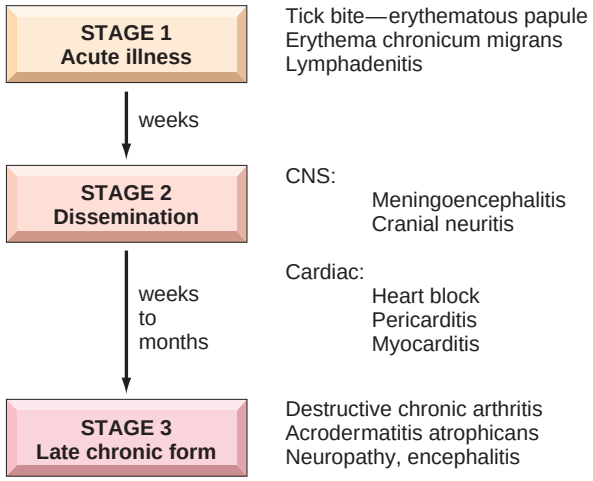

Figure 8-37 Clinical stages of Lyme disease.