DiPaola, 2009; Sharathkumar and Pipe, 2008). Although the symptomatology is similar regardless of which clotting factor is deficient, the identification of specific factor deficiencies allows definitive treatment with replacement agents.

In about 80% of all cases of hemophilia, the inheritance pattern is demonstrated as X-linked recessive. The two most common forms of the disorder are **factor VIII deficiency** (hemophilia A, or classic hemophilia) and **factor IX deficiency** (hemophilia B, or Christmas disease) with prevalence of approximately 1 in 5000 and 1 in 20,000 to 30,000 live births, respectively (McLean, Fiebelkorn, Temte, et al, 2013; Sharathkumar and Carcao, 2011; Zimmerman and Valentino, 2013). **Von Willebrand disease (vWD)** is another hereditary bleeding disorder characterized by a deficiency, abnormality, or absence of the protein called *von Willebrand factor (vWF)*. The following discussion is primarily concerned with factor VIII deficiency, which accounts for 80% of all hemophilia cases.

Pathophysiology

The basic defect of hemophilia A is a deficiency of **factor VIII (antihemophilic factor [AHF])**. Factor VIII is produced by the liver and is necessary for the formation of thromboplastin in phase I of blood coagulation (Fig. 24-4). The less factor VIII that is found in the blood, the more severe the disease. Individuals with hemophilia have two of the three factors required for coagulation: vascular influence and platelets. Therefore, they may bleed for longer periods but not at a faster rate.