is an important consideration in families in which female offspring may have inherited the trait.

Therapeutic Management

The primary therapy for hemophilia is replacement of the missing clotting factor. The products available are **factor VIII concentrates**, either produced through genetically engineering (recombinant form) or derived from pooled plasma, which are reconstituted with sterile water immediately before use. A synthetic form of vasopressin, **1-deamino-8-d-arginine vasopressin (DDAVP)**, increases plasma factor VIII activity and is the treatment of choice in mild hemophilia and vWD types I and IIA only if the child shows an appropriate response. After DDAVP administration, a threefold to fourfold rise in factor VIII level activity should occur. It is not effective in the treatment of severe hemophilia A, severe vWD, or any form of hemophilia B. Aggressive factor concentrate replacement therapy is initiated to prevent chronic crippling effects from joint bleeding.

Other drugs may be included in the therapy plan, depending on the source of the hemorrhage. Corticosteroids are given for hematuria, acute hemarthrosis, and chronic synovitis. Nonsteroidal antiinflammatory drugs (NSAIDs), such as ibuprofen, are effective in relieving pain caused by synovitis; however, they are occasionally used with caution because they inhibit platelet function (Curry, 2004; Hermans, De Moerloose, Fisher, et al, 2011). Oral administration of ε-aminocaproic acid (Amicar) prevents clot destruction. Its use is limited to mouth trauma or surgery with a dose of factor concentrate given first.

A regular program of exercise and physical therapy is an important aspect of management. Physical activity within reasonable limits strengthens muscles around joints and may decrease the number of spontaneous bleeding episodes.

Treatment without delay results in more rapid recovery and a decreased likelihood of complications; therefore, most children are treated at home. The family is taught the technique of venipuncture and to administer the AHF to children older than 2 to 3 years old. The child learns the procedure for self-administration at 8 to 12 years old. Home treatment is highly successful, and the rewards, in addition to the immediacy, are less disruption of family life, fewer