Nursing Care Management

The care of the child with AA is similar to that of the child with leukemia (see Chapter 25) and includes preparing the child and family for the diagnostic and therapeutic procedures, preventing complications from the severe pancytopenia, and emotionally supporting them in the face of a potentially fatal outcome. Information and support are available from the Aplastic Anemia and MDS International Foundation, Inc.*

The aspects of nursing care are discussed in the section on leukemia (see Chapter 25), therefore only interventions specific to AA are presented here. The drug ATG is usually administered by way of a central vein. If not, vigilant care must be directed to the IV infusion to prevent extravasation. Meticulous care of the venous access is essential because of the child's susceptibility to infection. Chemotherapeutic agents have been used in the treatment of relapsed patients with AA after unresponsive IST. Many of the side effects associated with chemotherapy such as nausea and vomiting, alopecia, and mucositis are experienced by children receiving treatment for AA. Specialized care is required for AA children who have HSCT that is discussed in Chapter 25.

Defects in Hemostasis

Hemostasis is the process that stops bleeding when a blood vessel is injured. Vascular and plasma clotting factors, as well as platelets, are required. A complex system of clotting, anticlotting, and clot breakdown (**fibrinolysis**) mechanisms exists in equilibrium to ensure clot formation only in the presence of blood vessel injury and to limit the clotting process to the site of vessel wall injury. Dysfunction in these systems leads to bleeding or abnormal clotting. Although the coagulation process is complex, clotting depends on three factors: (1) vascular influence, (2) platelet role, and (3) clotting factors.

Hemophilia

The term **hemophilia** refers to a group of bleeding disorders resulting from congenital deficiency or dysfunction or absence of specific coagulation proteins or factors (Montgomery, Gill, and