

vision. Chronic pulmonary disease, sinusitis, and otitis media result from repeated infections. In children who survive the bleeding episodes and overwhelming infections, malignancy presents an additional risk to survival. Medical treatment involves:

- Counteracting the bleeding tendencies with platelet transfusions
- Administering IVIG to provide passive immunity
- Administering prophylactic antibiotics to prevent and control infection
- Providing aggressive local therapy for the eczema

WAS is usually cured with HSCT and should be performed as early as possible ([Albert, Notarangelo and Ochs, 2011](#); [Buckley, 2011](#); [Mahlaoui, Pellier, Mignot, et al, 2013](#)). Several clinical trials focused on replacing the WAS gene are being conducted to determine the most effective vector ([Albert, Notarangelo, and Ochs, 2011](#)).

Nursing Care Management

Because of the poor prognosis for these children, the main nursing consideration is supporting the family in the care of a fatally ill child (see [Chapter 17](#)). Physical care should be directed at controlling the problems imposed by the disorder. The measures used to control bleeding are similar to those for hemophilia and vWD (see previous discussions). Another major goal is prevention or control of infection. Because eczema is a troublesome problem, nursing measures specific to this condition are especially important. The genetic implications of this X-linked recessive disorder differ little from those of any other X-linked disease.

Technologic Management of Hematologic and Immunologic Disorders

Blood Transfusion Therapy

Technologic advances in blood banking and transfusion medicine enable the administration of only the blood component needed by