

The definitive treatment for SCID is HSCT. If the condition is diagnosed at birth or within the first 3 months of life, more than 95% of cases can be treated successfully with HLA-identical or T-cell depleted haploidentical donor (usually a parent), or a matched unrelated donor bone marrow stem cells transplant (Bonilla and Geha, 2009; Buckley, 2011). Other approaches to management of SCID include providing passive immunity with IVIG infusions and maintaining child in a sterile environment. PCP prophylaxis is used to augment the humoral immunity until the transplant is performed. Several investigators are attempting gene therapy with some success, offering hope that gene therapy may eventually be the treatment of choice for cases of SCID (Bonilla and Geha, 2009; Buckley, 2011).

Nursing Care Management

Nursing care focuses on preventing infection and supporting the child and family. The care is consistent with that needed for HSCT for any condition (see earlier in this chapter). Because the prognosis for SCID is very poor if a compatible bone marrow donor is not available, nursing care is directed at supporting the family in caring for a child with a life-threatening illness (see Chapter 17). Genetic counseling is essential because of the modes of transmission in either form of the disorder.

Wiskott-Aldrich Syndrome

WAS is a congenital X-linked recessive disorder characterized by a triad of abnormalities: thrombocytopenia, eczema, and immunodeficiency of selective functions of B lymphocytes and T lymphocytes. An abnormal gene has been identified on the proximal arm of the X chromosome and designated the WAS protein (Bonilla and Geha, 2009; Buckley, 2011). At birth, the presenting feature may be increased bleeding at the circumcision site or bloody diarrhea as a result of thrombocytopenia. As the child grows older, recurrent infection and eczema become more severe, and the bleeding becomes less frequent.

Eczema is typical of the allergic type and readily becomes superinfected. Chronic infection with herpes simplex is a frequent problem and may lead to chronic keratitis of the eye with loss of