

successful bile drainage, many children ultimately develop liver failure and require liver transplantation.

Advances in surgical techniques and the use of immunosuppressive and antifungal drugs have improved the success of transplantation to survival rates of 80% to 90% ([Baumann and Ure, 2012](#)). The major obstacle continues to be a shortage of suitable infant donors.

## **Nursing Care Management**

Nursing interventions for the child with BA include support of the family before, during, and after surgical procedures and education regarding the treatment plan. In the postoperative period of a hepatic portoenterostomy, nursing care is similar to that following any major abdominal surgery. Teaching includes the proper administration of medications. Administration of nutritional therapy, including special formulas, vitamin and mineral supplements, gastrostomy feedings, or parenteral nutrition, is an essential nursing responsibility. Growth failure in such infants is common, and increased metabolic needs combined with ascites, pruritus, and nutritional anorexia constitute a challenge for care. The nurse teaches caregivers how to monitor and administer nutritional therapy in the home. Pruritus may be a significant problem that is addressed by drug therapy or comfort measures such as baths in colloidal oatmeal compounds and trimming of fingernails. The risk of complications of BA, such as cholangitis, portal hypertension, GI bleeding, and ascites, should be explained to the caregivers.

These children and their families require special psychosocial support. The uncertain prognosis, discomfort, and waiting for transplantation produce considerable stress. In addition, extended hospitalizations, pharmacologic therapy, and nutritional therapy can impose significant financial burdens on the family, as with any chronic condition. The Children's Liver Association for Support Services\* and the American Liver Foundation<sup>†</sup> provide educational materials, programs, support systems for parents of children with liver disease.

## **Structural Defects**