(Hassan and Balistreri, 2016). The definitive diagnosis of BA is further established during an exploratory laparotomy and an intraoperative cholangiogram that demonstrates complete obstruction at some level of the biliary tree.

Therapeutic Management

The primary surgical treatment of BA is hepatic portoenterostomy (Kasai procedure) in which a segment of intestine is anastomosed to the resected porta hepatis to attempt bile drainage. A Roux-en-Y jejunal limb is then anastomosed to the porta hepatis (a Y-shaped anastomosis performed to provide bile drainage without reflux). After the Kasai procedure, approximately one third of infants become jaundice free and regain normal liver function. Another one third of infants demonstrate liver damage; however, they may be supported by medical and nutritional interventions. A final third require liver transplantation.

Medical management of BA is primarily supportive. It includes nutritional support with infant formulas that contain medium-chain triglycerides and essential fatty acids. Supplementation with fat-soluble vitamins (A, D, E, and K); a multivitamin; and minerals, including iron, zinc, and selenium, is usually required. Aggressive nutritional support in the form of continuous gastrostomy feedings or TPN may be indicated for moderate to severe growth failure; the enteral solution should be low in sodium. Phenobarbital may be prescribed after hepatic portoenterostomy to stimulate bile flow, and ursodeoxycholic acid may be used to decrease cholestasis and the intense pruritus from jaundice. In cases of advanced liver dysfunction, management is the same as in infants with cirrhosis.

Prognosis

Untreated BA results in progressive cirrhosis and death in most children by 10 years old (Baumann and Ure, 2012). The Kasai procedure improves the prognosis but is not a cure. Biliary drainage can often be achieved if the surgery is done before the intrahepatic bile ducts are destroyed, and the success rate decreases to 20% if surgery is performed in an infant greater than 3 months old (Baumann and Ure, 2012). Long-term survival rates of 75% to 90% have been reported in children who receive the Kasai procedure (Baumann and Ure, 2012). However, even with