Cirrhosis

Cirrhosis occurs at the end stage of many chronic liver diseases, including BA and chronic hepatitis. Infectious, autoimmune, toxic injury, and chronic diseases such as hemophilia and cystic fibrosis can cause severe liver damage. A cirrhotic liver is irreversibly damaged.

Clinical manifestations of cirrhosis include jaundice, poor growth, anorexia, muscle weakness, and lethargy. Ascites, edema, GI bleeding, anemia, and abdominal pain may be present in children with impaired intrahepatic blood flow. Pulmonary function may be impaired because of pressure against the diaphragm caused by hepatosplenomegaly and ascites. Dyspnea and cyanosis may occur, especially on exertion. Intrapulmonary arteriovenous shunts may develop, which can also cause hypoxemia. Spider angiomas and prominent blood vessels on the upper torso are often present.

Diagnostic Evaluation

The diagnosis of cirrhosis is based on (1) the history, especially in regard to prior liver disease, such as hepatitis; (2) physical examination, particularly hepatosplenomegaly; (3) laboratory evaluation, especially LFTs such as bilirubin and transaminases, ammonia, albumin, cholesterol, and prothrombin time; and (4) liver biopsy for characteristic changes. Doppler ultrasonography of the liver and spleen is useful to confirm ascites, to evaluate blood flow through the liver and spleen, and to determine patency and size of the portal vein if liver transplantation is considered.

Therapeutic Management

Unfortunately, there is no successful treatment to arrest the progression of cirrhosis. The goals of management include monitoring liver function and managing specific complications such as esophageal varices and malnutrition. Assessment of the child's degree of liver dysfunction is important so that the child can be evaluated for transplantation at the appropriate time.

Liver transplantation has improved the prognosis substantially for many children with cirrhosis. The combination of new immunosuppressive medications and new surgical techniques has