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CURRENT PEDIATRIC THERAPY

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Hepatitis B related active hepatitis is more indolent and treatment is not so urgent. If the patient is hepatitis e antigen positive, corticosteroids should on no account be given as they increase virus replication. Treatment with antiviral drugs such as interferons must be considered, but their position has not been established and they are not generally available. If the patient is hepatitis e antigen negative, a trial of prednisolone may be considered, but if clinical and biochemical improvement has not followed three months' therapy, it should be stopped.

Disorders of the Hepatobiliary Tree

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Biliary Atresia, Biliary Hypoplasia, and Arteriohepatic Dysplasia (Alagilles Syndrome). Extrahepatic biliary atresia is responsible for approximately one-third of the cases of neonatal cholestatic jaundice. In 10% of biliary atresias, the lesion is either a distal atresia with patent proximal hepatic ducts or a cystic dilatation of ducts adjacent to the hilum of the liver. In these correctable forms of biliary atresia, reconstruction can be achieved by anastomosis of the patent extrahepatic bile duct or the gallbladder to a Roux-en-Y jejunostomy. Prognosis is excellent after biliary reconstruction.

In more common, noncorrectable forms of biliary atresia, there are no patent extrahepatic ducts, and the gallbladder is absent or rudimentary. For these lesions, hepatic portoenterostomy (Kasai operation) has been tried with varying degrees of success. Successful bile flow may be established in nearly all patients when the intrahepatic bile ducts at the hilum are greater than 150 μ in diameter but in only one-tenth of patients in whom such ducts are not demonstrable. In addition, the surgical success rate is related to the timing of operation. Success rates of 80–90% are reported if surgery is performed before 2 months of life, but the success rate falls to 20% after 3 months. It is deceptive to define success in terms of bile flow, since only 25% of patients are alive and free of jaundice 1–6 years after operation.

Cholangitis is a major problem after hepatic portoenterostomy. Prophylactic use of oral antibiotics, such as trimethoprim-sulfamethoxazole and cephradine, for a year has been advocated to suppress recurrent cholangitis. Various types of cutaneous enterostomy and complicated enteroenterostomy have been devised to reduce the risk of cholangitis, but none has been proved to be superior to a simple Roux-en-Y jejunostomy. Bleeding at or near the mucocutaneous junction is a common and serious complication of stomas that are left in place in the late stage of biliary atresia.

Revision of hepatic portoenterostomy should be limited to patients who became jaundice-free after an initial Kasai operation and who later develop recurrent jaundice due to obstruction or to a patient whose operation is judged to have been inadequate after review of previous medical records and pathology specimens.

Liver transplantation has become an alternative treatment of biliary atresia in the last several years. Before 1980, 1-year survival after liver transplantation for biliary atresia was 25% using conventional immunosuppressive therapy (azathioprine, prednisone, and antilymphocyte globulin). Since early 1980, 1-year survival has increased to 75%. Improvement has been due to better immunosuppression with cyclosporine and low doses of steroids. Five-year survival is projected at more than 50%.

Almost all recipients with biliary atresia have had earlier attempts at hepatic portoenterostomy. The 1-year survival rate of patients with biliary atresia is 20% less than that of patients with liver-based inborn metabolic errors, such as alpha$_1$-antitrypsin deficiency, tyrosinemia, and Wilson's disease. The previous hepatic portoenterostomy, which made the transplantation operation extremely difficult in many patients, has been responsible for the poorer outlook. Revisions and re-revisions of hepatic portoenterostomy, the placement of cutaneous enterostomy stomas, and the use of complicated enteric anastomoses other than simple Roux-en-Y jejunostomy are all adverse factors if liver transplantation is finally needed.

Although some authors advocate aggressive surgical therapy for biliary hypoplasia, hepatic portoenterostomy cannot be recommended. It is not only ineffective, but it also causes recurrent cholangitis. Arteriohepatic dysplasia (Alagilles syndrome) can be diagnosed by the characteristic features of broad forehead, deeply set and widely spread eyes, pointed chin, vertebral arch defects, pulmonary artery stenosis, and cholestatic jaundice due to a paucity of intralobular ducts. Both biliary hypoplasia and arteriohepatic dysplasia are good indications for liver transplantation.

Nutritional support is essential for children with these diseases to maintain their development. Vitamins, particularly A, D, and E, and calcium should be supplemented. Nutritional support consists of a diet high in proteins and low in fats. Formula with medium-chain triglycerides is now available.

Choledochal Cyst. In the past, choledochal cysts were drained by choledochocyst-duodenostomy or choledochocyst-jejunostomy in Roux-en-Y. The majority of patients so treated became relatively asymptomatic for a number of years after the

internal drainage operation, but many developed delayed complications, such as recurrent cholangitis, formation of biliary calculi, and progressive hepatic fibrosis. The development of malignant disease within the choledochal cysts or elsewhere in the biliary duct system has been reported repeatedly. The incidence of this complication is approximately 5%.

As a consequence of these complications and their increasing recognition, more and more surgeons are advising partial or complete excision of the cysts with performance of a Roux-en-Y jejunal anastomosis to the relatively normal proximal end of the duct system. If a part of the cyst wall cannot be resected because of technical difficulties, the mucosa lining should be stripped.

Caroli's Disease. Congenital segmental dilatation of the intrahepatic biliary duct system is called Caroli's disease. This disease is occasionally associated with congenital hepatic fibrosis, cystic spongiosis of the renal medulla, and cystic disease of the pancreas.

The large intrahepatic ducts of both lobes of the liver are usually involved. If the right and left main intrahepatic ducts are dilated at the hilum, a large hepaticojejunostomy in Roux-en-Y will relieve bile stasis, thereby reducing the chance of cholangitis and stone formation. If many biliary stones have already formed, the tip of the jejunal Roux loop is brought up to the skin to create the jejunal stoma. Through this stoma, the retained stones can be removed postoperatively by endoscopic or radiologic maneuvers.

Rarely, the cystic dilations are confined to one lobe of the liver. Hepatic resection is indicated only in these unusual circumstances.

Cholecystitis and Cholelithiasis. Cholelithiasis in children is usually a complication of hemolytic diseases, such as hereditary spherocytosis, sickle cell disease, and thalassemia major. Adult types of cholesterol stones are occasionally seen in postpubertal children. Children with cholelithiasis should undergo elective cholecystectomy in the same way as adults. If the cholelithiasis is complicated by acute cholecystitis, the child should be treated with antibiotics first. If the signs of acute inflammation subside with antibiotic therapy, the cholecystectomy is performed electively during the same hospitalization, but if there is not a good response within 48–72 hours, emergency cholecystectomy is carried out.

Acalculous cholecystitis may occur in children during acute febrile illnesses, severe diarrhea, and postoperative convalescence, or it may follow major trauma and large burns. These patients are ordinarily treated by intensive antibiotic therapy and gastric suction. However, if gallbladder distension is progressive, cholecystectomy is performed. If recovery from the acute illness is complete, residual gallbladder disease is not apparent.

Pancreatic Diseases

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ACUTE PANCREATITIS

The general principles of treatment of acute pancreatitis are (1) to treat hypovolemia and electrolyte abnormalities, (2) to relieve pain, (3) to reduce pancreatic secretions, and (4) to remove the precipitating cause.

Correction of hypovolemia should begin immediately, utilizing a large-bore central venous catheter for fluid replacement and to monitor central venous pressure. Hypotension and low central venous pressure should be corrected as rapidly as possible with plasma, dextran, albumin, or whole blood. Shock is the main cause of death in acute pancreatitis. Shock is primarily a result of exudation of plasma into the retroperitoneal space and peripheral vasodilatation caused by increased kinin activity.

After hypovolemia has been corrected, the rate of intravenous infusions should be reduced so as to provide maintenance plus replacement of ongoing losses from nasogastric suctioning and exudation into the peritoneal and retroperitoneal spaces. Monitoring urine output and central venous pressure are mechanisms for assessing the adequacy of the fluid replacement. Major complications of treatment of severe acute pancreatitis are pulmonary edema and congestive heart failure; these usually occur 3 to 7 days after the onset of pancreatitis. Though in many cases the cause is unknown, in some cases fluid overload has occurred because of excessive fluid replacement. Thus, the amount of fluid replacement must be adjusted frequently for changes in intravascular volume.

Serum electrolytes, including calcium and magnesium, serum creatinine, and blood urea nitrogen determinations, will aid in selecting the appropriate electrolyte composition of intravenous solutions. Since between 2 and 17 per cent of patients with acute pancreatitis have renal failure, potassium should not be added to IV solutions until stable urine output has been established. In addition to maintenance sodium chloride and potassium chloride of 3 mEq/kg/24 hr and 2 mEq/kg/24 hr, respectively, losses from nasogastric suctioning should be replaced. Though 5 per cent dextrose solutions should be initiated, hyperglycemia and hypoglycemia occasionally seen in severe pancreatitis warrant careful monitoring of urinary reducing substances and blood glucose