

Treatment

Treatment

For breast-fed infants, introducing supplemental formula feeds using a medium-chain triglyceride-based feed and fat-soluble vitamin supplementation (titrated according to growth) is a priority . Patent segments of proximal bile duct are found in 10% of type I lesions. A direct Roux-en-Y hepaticojejunostomy Daniel Alagille , 1925–2005, paediatric hepatologist, Hôpital Bicêtre, Paris, France. César Roux , 1857–1934, Professor of Surgery and Gynaecology , Lausanne, Switzerland, described the Roux-en-Y loop in 1908. Jacques Caroli , 1902–1979, gastroenterologist, Hôpital St Antoine, Paris, France, described cavernous ectasia in the biliary tree in 1958. will achieve bile flow in 75%, but progressive fibrosis r esults in disappointing long-term results. A simple biliary-enteric anas - tomosis is not possible in the majority of cases in which the proximal hepatic ducts are either very small (type II) or atr etic (type III). These are treated by the Kasai procedure , in which radical excision of all bile duct tissue up to the liver capsule is performed. A Roux-en-Y loop of jejunum is anastomosed to the exposed area of liver capsule above the bifurcation of - the portal vein, creating a portoenterostomy . The chances of achieving e ff ective bile drainage after portoenterostomy are maximal when the operation is performed before the age of 8 weeks, and approximately 90% of children whose bilirubin falls to normal can be expected to survive for 10 years or more. Early referral for surgery is critical. Postoperative complications include bacterial cholangi - tis, which occurs in 40% of patients. Repeated attacks lead to hepatic fibrosis, and 50% of long-term survivors develop portal hypertension, with one-third having variceal bleeding. Liver transplanta tion should be considered in children in whom a portoenterostomy is unsuccessful. Results are improv - ing, with 70–80% alive 2–5 years following transplant.

Atretic Patent Atretic IIb III

Treatment

Asymptomatic gallstones do not need intervention, however prophylactic cholecystectomy may be performed for asymp - tomatic cholelithiasis in the following situations: /uni25CF large (>3 /uni00A0 cm) gallstones; /uni25CF choledocholithiasis; /uni25CF chronic haemolytic conditions (sickle cell disease, heredi - tary spherocytosis); /uni25CF gallbladder polyps >1 /uni00A0 cm in diameter; /uni25CF suspicion/risk of malignancy (anomalous pancreatic duc - tal drainage); /uni25CF calcification of the wall (porcelain gallbladder); /uni25CF some ethnic groups or subjects living in areas with a high prevalence of gallbladder cancer associated with gallstones (some parts of northern India, Native Americans, Mexican Americans, Colombia, Chile, Bolivia); /uni25CF transplant patients (during transplantation); /uni25CF bariatric surgery . For patients with symptomatic gallstones, cholecystectomy is the treatment of choice if there are no medical contraindi - cations. The initial non-operative treatment is based on four steps: 1 Nil by mouth and intravenous fluid administration until the pain resolves. 2 Analgesics. 3 Antibiotics. As the cystic duct is blocked in most instances, the concentration of antibiotic in the serum is more import - ant

than the concentration in the bile. A broad-spectrum antibiotic effective against Gram-negative aerobes is most appropriate (e.g. cefazolin, cefuroxime or ciprofloxacin). 4 Subsequent management. When the temperature, pulse and other physical signs show that the inflammation is subsiding, oral fluids are reinstated, followed by a regular diet. If jaundice with deranged ALP and enzyme levels is present, MRCP should be performed to exclude choledocholithiasis. If there is any concern regarding the diagnosis or the presence of complications such as perforation, CT should also be performed. The timing of surgery in acute cholecystitis remains controversial. Early cholecystectomy, undertaken by an experienced surgeon with excellent operating facilities within 5–7 days of the onset of the attack, is safe and shortens total hospital stay. Nevertheless, the conversion rate in laparoscopic cholecystectomy is higher in acute than in elective surgery. If early operation is not indicated, one should wait approximately 6 weeks for the inflammation to subside before operating. The Tokyo Guidelines (2013/2018) allow the assessment of severity and grading of acute cholecystitis (Table 71.2) and provide a consensus-derived treatment algorithm based on grading, patient comorbidity and the facilities and expertise available (Figure 71.26). et al 25 Acute and chronic inflammation of the gallbladder can occur in the absence of stones and give rise to a clinical picture similar to that of calculous cholecystitis. Some patients have non-specific inflammation of the gallbladder, whereas others have one of the cholecystoses. Acute acalculous cholecystitis is particularly seen in critically ill patients and those recovering from major surgery, trauma and burns. The diagnosis is often missed and the mortality rate is high. The treatment is cholecystectomy for patients who are able to tolerate surgery. In selected patients, non-surgical treatment (such as antibiotics or percutaneous cholecystostomy) may be an effective alternative to surgery.

TABLE 71.2 Tokyo Consensus Guidelines for severity grading of acute cholecystitis. Grade III (severe) acute cholecystitis Associated with dysfunction of any one of the following organs/systems: 1 Cardiovascular dysfunction Hypotension requiring treatment with dopamine ≥ 5 $\mu\text{g}/\text{kg}/\text{min}$, or any dose of epinephrine 2 Neurological dysfunction Decreased level of consciousness 3 Respiratory dysfunction $\text{PaO}_2/\text{FIO}_2$ ratio <300 4 Renal dysfunction Oliguria; creatinine >2.0 mg/dL 5 Hepatic dysfunction Prothrombin time (PT-INR) >1.5 6 Haematological dysfunction Platelet count $<100,000/\text{mm}^3$ Grade II (moderate) acute cholecystitis Associated with any one of the following conditions: 1 Elevated white cell count ($>18,000/\text{mm}^3$) 2 Palpable tender mass in the right upper abdominal quadrant 3 Duration of complaint >72 hours 4 Marked local inflammation (gangrenous cholecystitis, pericholecystic abscess, hepatic abscess, biliary peritonitis, emphysematous cholecystitis) Grade I (mild) acute cholecystitis Does not meet the criteria of grade II or grade III acute cholecystitis. Grade I can also be defined as acute cholecystitis in a healthy person with no organ dysfunction and mild inflammatory changes in the gallbladder, making cholecystectomy a safe and low-risk operative procedure $\text{PaO}_2/\text{FIO}_2$ ratio is the ratio of arterial oxygen partial pressure (PaO_2 in mmHg) to fractional inspired oxygen (FIO_2) expressed as a fraction (not a percentage) at sea level, the normal $\text{PaO}_2/\text{FIO}_2$ ratio is $\sim 400\text{--}500$ mmHg ($\sim 55\text{--}65$ kPa); PT-INR, prothrombin time–international normalised ratio. Reproduced with permission from Yokoe M. Tokyo Guidelines 2018: diagnostic criteria and severity grading of acute cholecystitis (with videos). *J Hepatobiliary Pancreat Sci* 2018; (1) 41–54.

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