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with appropriate antibiotics, and cholecystectomy may be necessary if the gallbladder fails to reduce in size.

PRIMARY SCLEROSING CHOLANGITIS

Primary sclerosing cholangitis (PSC) is a chronic disorder of the larger bile ducts. It may involve any portion of the intrahepatic or extrahepatic biliary system and results in strictures, stone formation, bile stasis, and decreased liver synthetic function. Its etiology is unknown, and there is a higher frequency of HLA DR3 and B8 in these patients than in healthy individuals. The characteristic lesion of PSC is fibroobliterative bile duct destruction, but this is present in only 10% to 50% of patients undergoing liver biopsy. Thus, histologic diagnosis can be difficult. The primary role of liver biopsy, therefore, is to both exclude other diseases and confirm that the pathologic changes present are consistent with PSC. Endoscopic retrograde cholangiopancreatography (ERCP) is a highly sensitive test for demonstrating PSC. This disease occurs predominantly in males and is often associated with both ulcerative colitis and immunodeficiency states. It may be caused by an immune-mediated process, as portal infiltrates consist mainly of T cells, and there are circulating autoantibodies, of which those directed against neutrophils have sparked the most interest. These perinuclear antineutrophil cytoplasmic antibodies (pANCA) have been found also in a significant number of patients with ulcerative colitis. UDCA and immunosuppressive agents such as methotrexate are the principal agents used to achieve remission. Once complete obstruction occurs, therapeutic ERCP may permit dilatation of stenotic areas of the affected bile ducts. With disease that progresses despite maximal medical therapy, patients become candidates for OLT.

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TUMORS OF THE LIVER

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Increasing numbers of hepatic mass lesions are found incidentally by advanced imaging technology. Although most of these incidental tumors are histologically benign and do not require any therapy, they must be thoroughly investigated. Modern imaging technology is quite efficient in detecting small lesions but is not effective in producing pathognomonic findings of many hepatic lesions other than hemangiomas and cysts. Percutaneous needle biopsy often fails to establish a definitive diagnosis because of its limited sampling, and it can cause serious hemorrhage when unwisely performed for vascular lesions.

Major hepatic resections can now be performed with minimum operative risk (less than 5%), but no surgeon should explore a hepatic mass without having the competence to perform all of the major resections, including a right and left trisegmentectomy.

BENIGN TUMORS

Most of the benign tumors of the liver are asymptomatic and are found incidentally during studies for other disorders or during abdominal operations. The general approach to a small incidental tumor (less than 3 cm in diameter) that is considered benign is close observation after thorough investigation. When the tumor changes its imaging characteristics or increases in size during close observation, it must be excised immediately. Larger incidental tumors, other than asymptomatic cavernous hemangiomas, deserve excisional therapy unless unequivocal benignity is confirmed.

Hemangiomas are the most common benign tumors of the liver. Giant cavernous hemangiomas should be treated by surgical excision, particularly when they are symptomatic (e.g., pain, mass-related complaints) or are found to have a necrotic center inside. The majority of giant cavernous hemangiomas require lobectomies or trisegmentectomies of the liver, but some located on the surface of the liver or pedunculated can be enucleated along pseudocapsular margins without significant loss of normal liver tissue. Ligation or embolization of the feeding hepatic artery and radiation therapy may be hazardous and do not have long-standing effects on the course of giant cavernous hemangiomas.

Infantile hemangioendotheliomas are seen most often in infants during the first 6 months of life and are distinct from cavernous hemangiomas. The lesions should be excised by anatomic hepatic resection whenever possible. Treatment with prednisone, diuretics, and digoxin can be used initially when the patient's condition prohibits surgery or the lesion is too extensive for resection. Response to prednisone may allow surgery to be performed safely in a few weeks. In extensive lesions, radiation to the liver may be used after pathologic diagnosis is confirmed. Favorable responses to steroids, radiation, and hepatic artery ligation or embolization have been reported. The treatment should be vigorous because complete regression and cure are possible.

Other benign tumors include liver cell adenoma, focal nodular hyperplasia, hematoma, mesenchymoma, teratoma, and fibroma. Radiologic differentiation of these benign tumors from malignant tumors is unreliable. Pathologic confirmation of benign tumors is mandatory for each lesion. Large benign tumors should be treated by surgical excision, particularly when they are symptomatic. Adenoma has a tendency to rupture and cause life-threatening hemorrhage. Some adenomas cannot be easily differentiated from low-grade hepatocellular carcinoma by needle biopsies. If the diagnosis is uncertain, the lesion should be excised with an adequate margin without delay.

Congenital hepatic cysts are usually asymptomatic and do not require any therapy. Although aspiration, internal drainage, marsupialization, fenestration, and sclerotherapy have all been recommended for symptomatic congenital cysts, these approaches are no longer justifiable for the treatment of single or localized multiple cysts because hepatic resections can be performed quite safely now.

MALIGNANT TUMORS

The most common primary malignant tumor of the liver in children is hepatoblastoma. Hepatocellular carcinoma is the second most common and usually occurs in older children. Sarcomas of the liver, such as rhabdomyosarcoma and angiosarcoma, are rare. None of these has a favorable outlook, but fibrolamellar hepatocellular carcinoma, which is common in older children and young adults, has a better prognosis than other types of malignancy.

The treatment for all malignant liver tumors is complete surgical excision by anatomic hepatic resection. Hepatic resection of more than the right or left lobe of the liver can be performed quite safely. For example, a large tumor occupying the right lobe of the liver and the medial segment of the left lobe can be resected by right hepatic trisegmentectomy, leaving only the left lateral segment of the left lobe (to the left of the falciform ligament), or a large tumor occupying the left lobe and the anterior segment of the right lobe can be resected by left hepatic trisegmentectomy, leaving only the posterior segment of the right lobe (posterior to the right hepatic vein). These major hepatic resections can now be performed by experienced surgeons with less than a 5% operative mortality.

We have found that computed tomography scan or magnetic resonance imaging is most useful in assessing the extent of the tumor, but findings can be misleading, particularly when a large tumor distorts normal anatomic boundaries. If the resectability is uncertain after extensive preoperative investigation, the patient should be referred to

a surgeon who is experienced in major hepatic resection rather than undergo exploratory celiotomy by someone who is unprepared to undertake a definitive procedure.

After curative hepatic resection, we usually recommend that patients receive adjuvant chemotherapy for at least 1 year. We have been using combination chemotherapy with doxorubicin, dactinomycin, vincristine, and cyclophosphamide, and often mitomycin or cisplatin. The value of this approach has not been validated in randomized trials, but the patients who have received adjuvant chemotherapy after curative resections of large tumors have seemed to have longer tumor-free survival.

In general, liver transplantation (total hepatectomy and liver replacement) cannot offer good long-term results when applied to large malignant tumors that cannot be removed by subtotal hepatectomy. However, liver transplantation can result in a cure (more than 5-year survival) on more than isolated occasions. The most favorable lesions for transplantation, just as with resection, are the fibrolamellar hepatoma and epithelioid hemagioendothelial sarcoma. On the other hand, most of the patients who have received liver transplantation for other end-stage liver diseases, such as tyrosinemia and alpha$_1$-antitrypsin deficiency disease, and whose malignant tumors were small and incidental survived tumor free for several years.

The most common metastatic liver tumors in children are neuroblastoma and Wilms' tumor. Although chemotherapy and radiation therapy may be helpful in treating these metastatic tumors, the lesion should be excised whenever possible, particularly if it is localized to part of the liver. Hepatic resections for metastatic tumors are much safer than those for primary malignancy.

INTESTINAL TRANSPLANTATION

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The clinical entity of intestinal failure includes an extremely varied array of causative diseases, associated anomalies, and complicating factors, with one common denominator: they all rely on total parenteral nutrition (TPN) for their survival. The introduction of TPN has improved the outcome of many patients and is the accepted therapeutic modality today. However, some patients suffer complications of this therapy, principally liver dysfunction and venous access complications, that limit its long-term use. Also, the frequent morbidity and need for specialized care and hospitalizations place a significant drain on the health care system. Finally, the social awkwardness and emotional demands on the family can hinder tremendously the normal developmental and behavioral maturity of children. There is an urgent need for achieving success with intestinal transplantation.

INDICATIONS

The inability to maintain a normal nutritional status by the use of the gastrointestinal tract alone constitutes intestinal failure (IF). Children with IF usually have had major bowel resections because of catastrophic gastrointestinal disease or suffer functional abnormalities as a result of motility or secretory/absorptive disorders.

The following are **major indications for intestinal transplantation** in children:

- Congenital malformations
- Gastroschisis
- Hirschsprung's disease
- Necrotizing enterocolitis
- Pseudoobstruction
- Volvulus

The composition of the allograft type focuses on the integrity of the remaining gut and other abdominal organs. Those patients with IF and cholestatic liver disease secondary to TPN are candidates for combined liver intestinal transplant (LITX). Guidelines used in assessing the need for a concomitant liver transplant include biochemical parameters, histology, and the presence of portal hypertension. Patients with functional disorders may require replacement of the entire gastrointestinal tract. Candidates for isolated intestinal transplant (ITX) are those who present incipient signs of TPN injury to the liver, although without significant damage, and those who have suffered multiple septic episodes from line infection, with consequent thrombosis, and are limited in the availability of access sites. These patients are in danger of progressively depleting these sites and should be considered for ITX.[3]

A potential small bowel transplant recipient should have a thorough assessment of nutritional history and present nutritional status, as well as an outline of the abdominal anatomy. This should allow for planning of the optimum time for transplantation, as well as for accurate planning of graft type.

THE OPERATION

A history of normal intestinal function in a patient referred for potential liver donation is considered adequate for possible intestinal donation. The donor should be hemodynamically stable, without a need for vasopressor support. There should be no history of cardiac arrest. The adequacy of the intestine is assessed by the donor team at the time of surgery. We select a donor of similar or smaller size than the recipient. The ABO blood groups should be identical, and there is no HLA matching.

The procurement of multiple visceral organs (en-bloc or as separate components) focuses on the isolation and cooling of the organs, preserving their vascular and parenchymal anatomy. The complete multivisceral retrieval includes the stomach, duodenum, pancreas, liver, and small and large intestine. After encirclement of the proximal aorta near the diaphragm and the distal aorta below the origin of the inferior mesenteric artery, the organs are flushed with chilled University of Wisconsin (UW) solution via a catheter inserted into the lower abdominal aorta. In situations where the liver is not required in the graft, it may be separated in situ after perfusion or at the back table. Back table irrigation of the intestinal lumen is required only when the colon is included in the graft. Manipulation of the graft lymphoreticular tissue (using polyclonal or monoclonal antilymphocyte antibody preparations or radiation) is not performed.

The recipient operation focuses on preservation of the remaining bowel and the status of the native liver. The final judgment as to graft type is made at this time. The liver is usually excised, preserving the retrohepatic vena cava as for a piggyback transplant. A portacaval shunt is used when the liver is removed, and the proximal gut is preserved (stomach, duodenum, pancreas, spleen), thus allowing decompression of these organs. This is not required in recipients of a complete multivisceral transplant (MVTX) or ITX graft.

Revascularization in the MVTX or LITX graft uses a Carrel patch containing the celiac and superior mesenteric arteries, which is anastomosed to the recipient infrarenal aorta (with or without an interposition graft of donor thoracic or abdominal aorta). The venous drainage of the graft is into the hepatic veins of the recipient. After reperfusion, the portacaval shunt may be taken down, and a recipient portal vein-to-donor portal vein anastomosis may be performed. In the ITX graft, the superior mesenteric artery is anastomosed into the infrarenal aorta. The venous drainage of this type graft can be into the native superior mesenteric vein, the portal vein at the level of the hepatic hilus, or the inferior vena cava. The graft is reperfused after unclamping of the arterial inflow, allowing bleeding to occur from the venous outflow anastomotic lines. This allows for drainage of the potassium-rich UW solution. The venous drainage clamps are then removed. The gastrointestinal tract is reconstructed in a standard fashion, anastomosing to native intestine proximally and distally. Biliary reconstruction is required only in recipients of an LITX graft and is