Polycystic Liver Disease: Multimodality Imaging for Complications and Transplant Evaluation¹

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LEARNING OBJECTIVES

After reading this article and taking the test, the reader will be able to:

■ Identify patients with polycystic liver disease and list the different genetic mutations in patients with versus those without autosomal dominant polycystic kidney disease.

• Describe diagnostic imaging features of cysts and patterns of cyst size and distribution that affect surgical or other therapeutic decision making in patients with symptomatic polycystic liver disease.

• Discuss complications and expected outcomes of various therapies as well as appropriate imaging modalities for patients with symptomatic polycystic liver disease.

TEACHING POINTS

See last page

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Polycystic liver disease (PLD) is usually associated with polycystic kidney disease but may also occur as an isolated finding in a rarer genetically distinct disease. In either case, the cyst burden will progress over time and, in rare cases, may affect liver function or become symptomatic due to massive hepatomegaly. The character, distribution, location, and size of hepatic cysts are important. Computed tomography, magnetic resonance imaging, or ultrasonography may provide the surgeon with valuable preoperative information, such as the location of infected or hemorrhagic cysts that may be responsible for symptoms. Less invasive cyst aspiration or fenestration may provide temporary relief from dominant or symptomatic cysts, but these cysts will recur in up to 75% of patients. Cyst fenestration with partial hepatic resection and liver transplantation are two therapies that provide more permanent resolution of symptoms in patients with extensive hepatic involvement. However, the higher risk of complications associated with more aggressive surgical therapy must be considered when determining the appropriate therapy for a given patient. Knowledge of the cyst patterns and available treatment options in patients with PLD will help the radiologist provide the referring clinician with important information for therapeutic decision making.

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 $\label{eq:Abbreviations: ADPKD = autosomal dominant polycystic kidney disease, PCLD = autosomal dominant polycystic liver disease, PLD = polycystic liver disease$

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a.

b.

Figure 1. PLD in an asymptomatic 44-year-old woman undergoing screening for ADPKD. Coronal gadoliniumenhanced gradient-echo T1-weighted magnetic resonance (MR) image (a) and half Fourier acquisition single-shot turbo spin-echo T2-weighted MR image (b) demonstrate cysts of varying size throughout all segments of the liver.



a.

Figure 2. Symptomatic PLD and ADPKD. (a) Computed tomographic (CT) scan obtained with orally administered contrast material demonstrates polycystic liver and kidney disease, with multiple calcifications along the cyst wall in the left kidney. (b) CT scan obtained inferior to a reveals mass effect on the hepatic flexure of the colon (arrow). Many patients with PLD have renal insufficiency due to ADPKD. Mass effect can be well demonstrated at unenhanced CT; however, vascular anatomy and patency and biliary dilatation are not well depicted without intravenous contrast material in these patients.

Introduction

Polycystic liver disease (PLD) is a hereditary condition that may arise either in patients with autosomal dominant polycystic kidney disease (ADPKD) or in patients with a different genetic mutation that results solely in autosomal dominant polycystic liver disease (PCLD). Both

disorders are characterized by progressive development of fluid-filled biliary epithelial cysts throughout all segments of the liver. These cysts arise from malformation of the embryonic ductal plate, with formation of von Meyenburg complexes (hamartomas) that are lined with functional biliary epithelium (1). Most patients with PLD are asymptomatic (Fig 1). Despite impressive physical examination and radiologic findings, only a minority of patients with PLD will progress **Radio**Graphics



b.

Figure 3. PLD in a 47-year-old woman with massive abdominal distention due to hepatomegaly. (a, b) Frontal (a) and lateral (b) CT scanograms demonstrate a markedly protuberant abdomen. (c) Unenhanced CT scan demonstrates marked displacement of the stomach posteriorly (arrow). (d) Intravenous contrast material-enhanced CT scan obtained at the same level as c clearly depicts the right hepatic artery (arrow) replaced to the superior mesenteric artery. The patient suffered from early satiety and progressive immobility and required liver transplantation despite essentially normal liver function tests. The explanted liver weighed 10,190 g and measured $42 \times 40 \times 20$ cm. Of note, the patient had a normal creatinine level. Three of her siblings had already undergone kidney transplantation for ADPKD, but she was the only sibling with PLD.

to advanced liver disease or develop complications as a result of massive hepatomegaly (2,3). However, this minority of patients may become incapacitated by the disease, with symptoms of severe abdominal pain (Fig 2), massive abdominal distention (Fig 3), early satiety, supine dyspnea, and regurgitation. Most patients maintain normal liver function; however, cholestatic enzyme patterns, portal hypertension, cyst infection, or spontaneous and life-threatening intracystic hemorrhage may occasionally occur. These complications rarely occur in the absence of hepatomegaly. There is no known effective medical therapy (3).

| Therapy | Symptom Severity | Cyst Anatomy and Distribution | Complications | Effectiveness (Symptomatic Relief)* |
|---|--|---|--|--|
| Cyst aspiration, injection of a sclerosant | Mild | Best option for isolated large dominant cysts that are amenable to percutaneous needle insertion | Recurrence | Good in short term, recurrence rate nearly 100% in intermediate term |
| Cyst fenes- tration | Moderate to severe | Multiple large, superfi- cial cyst segments in hepatic segments II–VI; not a good option for posterior segments | Perioperative fluid loss; postoperative ascites, pleural effusions, bile leak, hemorrhage | Good in short term, moderate to good in intermediate term, recurrence in long term |
| Cyst enucle- ation | Moderate | Isolated or a few domi- nant cysts | Ascites, hemorrhage, bile leak | Good in short terms poor in interme- diate term, espe- cially in patients with ADPKD and PLD |
| Cyst fenestra- tion with par- tial hepatic resection | Severe | Multiple large cysts with regions of innu- merable smaller cysts, focal island of relatively spared pa- renchyma | Nonsegmental resection required due to ana- tomic distortion by cysts; perioperative fluid loss; postopera- tive ascites, pleural effusions, bile leak, hemorrhage | Good in short term, moderate to very good in interme- diate term, hy- pertrophy of spared paren- chyma in long term |
| Liver trans- plantation | Severe (refractory pain, debilitating abdominal dis- tention, "lethal exhaustion" syn- drome) | Massive hepatomegaly, any distribution | Adhesions from prior cyst, therapies hinder explantation, greater risk when combined with kidney trans- plantation, greater intraoperative blood loss, surgical mortal- ity rates of 10%–33% | Excellent in long term |

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Treatment options depend on symptoms, cyst anatomy, and cyst distribution (Table). Treatment for milder cases may include cyst aspiration and injection of a sclerosant such as ethanol. For more severely symptomatic individuals, therapeutic options include surgical (open or laparoscopic) cyst fenestration, cyst enucleation, or cyst fenestration combined with partial hepatic resection. Ultimately, some patients require liver transplantation for refractory symptoms. The trade-off between less invasive procedures that provide

immediate short-term relief and more invasive therapies that provide better long-term outcomes but are technically challenging due to the markedly distorted anatomy in these individuals must be considered for each individual case. In this article, we review genetic and anatomic considerations in PLD. In addition, we discuss and illustrate imaging findings, therapeutic options, and complications in patients with PLD.

Genetic Considerations

PLD is genetically heterogeneous. Most patients with PLD have ADPKD (Figs 1-3) (3). ADPKD



a.

Figure 4. PLD associated with ADPKD in a 56-year-old woman. The patient underwent combined liver transplantation, kidney transplantation, and right nephrectomy. (a) Unenhanced CT scan shows nearly complete replacement of the hepatic parenchyma by cysts. The explanted liver weighed 8285 g. (b) Photograph of the sectioned specimen shows sparing of a small portion of the right lobe (arrow), a finding that corresponds to the island of parenchyma seen at CT.

affects one in 500-1000 individuals and results from mutations on chromosomes 4 and 16. Hepatic involvement occurs in 30%-70% of patients with ADPKD (1,4-7), most of whom are female. The relationship between gender and cyst development is likely best explained in terms of hormonal influences: The prevalence of hepatic cysts is reported to be 58%-75% in female patients and 42%-62% in male patients with ADPKD (2). There is also a higher prevalence with increasing age, severity of renal cystic disease, and degree of renal dysfunction (8). As dialysis has become widely available, patients with ADPKD are surviving longer and developing hepatic complications of the disease (6,9). In the past, it has been difficult to differentiate between patients with ADPKD and hepatic involvement and those with PCLD because the liver pathology is indistinguishable in these two groups of patients (10). Although PCLD was first postulated in 1986 as a genetic disease distinct from known forms of ADPKD (11), only recently have the two genes responsible for PCLD been discovered: the PRKCSH gene, located on chromosome 19p; and the SPC 63 gene, located on chromosome 6q. These genes encode glycoproteins produced within the endoplasmic reticulum; mutations result in defective processing of a key regulator of biliary cell growth (12). The defect in encoding of biologic proteins by the genes for PCLD is different from the defect associated with ADPKD (13),

although the end result of all these mutations is the production of biliary-type cysts in the liver. PCLD is a milder disease than ADPKD and, unlike the latter, does not lower life expectancy. Renal cysts can occur, but there is no impairment of renal function (13). PCLD is much less common than ADPKD, with a prevalence of less than 0.01% based on autopsy data (2). In most reported series, the majority of patients treated for PLD have ADPKD.

Anatomic Considerations

The course of PLD is variable but progressive. In patients with ADPKD, the number and size of cysts increase with advancing age. Likewise, PCLD manifests as an enlarged, diffusely cystic liver (14). Hereditary polycystic livers typically contain more than 20 cysts, helping differentiate them from (nonhereditary) multicystic livers (12), and they generally demonstrate replacement of over 50% of the hepatic parenchyma by cysts (15). In other series of younger patients with ADPKD, an arbitrary number of cysts as low as four to six qualifies a patient as having a polycystic liver (1,2). However, symptomatic polycystic liver patients have massive hepatomegaly. Explanted polycystic livers reported in the literature weighed an average of 10 kg (range, 5-13 kg) (Fig 4) (9, 16,17), whereas normal livers typically



weigh 1.4–1.6 kg. The size and distribution of the cysts may elongate or compress critical vascular or ductal structures; pretransplantation evaluation with duplex Doppler ultrasonography (US) or CT with intravenous (creatinine-permitting) and oral contrast material help alert the transplantation surgeon to these conditions. MR imaging with MR cholangiopancreatography may be especially useful for delineating the ducts in these patients, in addition to clarifying the anatomic relationships of the cysts to critical vascular structures after gadolinium-based intravenous contrast material enhancement.

Imaging Findings

Cysts

Two types of cysts may be found in the livers of patients with PLD: intrahepatic cysts and peribiliary cysts (Fig 5). Intrahepatic cysts are more common, being found in 56 of 64 patients in one series (18) and in 84 of 93 patients in another (19). These cysts arise from von Meyenburg complexes and are mostly peripheral, ranging from less than 10 mm to 80 mm in size (7). Peribiliary cysts were reported in 41 of 56 patients (18) and 59 of 90 patients (19) in the two large imaging series mentioned earlier. These cysts arise from dilated peribiliary glands (2); are typically less than 10 mm in diameter; and appear as either discrete cysts, a string of cysts, or tubular structures paralleling the path of the portal vessels (7, 18, 19).

Identification of dominant cysts that may be amenable to temporizing percutaneous aspiration and sclerosis is helpful in symptomatic patients (Fig 6); this therapy is often repeated over a period of years before ultimately choosing transplantation as a therapeutic option. Although US, CT, and MR imaging may adequately demonstrate dominant cysts, signal intensity changes at MR imaging may best characterize cysts complicated by hemorrhage or infection that are producing acute worsening of symptoms (6). Increased



Figure 5. PLD in a 58-year-old man undergoing peritoneal dialysis for end-stage ADPKD. Intravenous contrast–enhanced CT scan through the hepatic hilum demonstrates enhancement of the right and left portal veins. There are multiple small peribiliary cysts (arrows) with a "string of pearls" distribution along the portal vessels. A few intrahepatic cysts are also present. No definite biliary dilatation is seen.

T1-weighted signal intensity at MR imaging (Fig. 6), heterogeneous echotexture at US, or increased attenuation at unenhanced CT may be seen with hemorrhage but are not specific, also being possible with cyst infection. CT signs of cyst infection (besides heterogeneous increased attenuation) include a fluid-fluid level in the cyst, cyst wall thickening or calcification, and intracystic gas bubbles (2). In one small series, fluorine 18 (¹⁸F) fluorodeoxyglucose positron emission tomography proved very helpful in both diagnosing and excluding renal and hepatic cyst infections. In this series, increased ¹⁸F-fluorodeoxyglucose uptake was seen in infected renal and hepatic cysts but was not seen when there was no infection (20).

Liver Size and Cyst Distribution

Imaging can be used to guide individual cyst therapies as well as procedures to ameliorate massive hepatomegaly. Cyst size and patterns of cyst distribution affect therapeutic options.

Teaching Point Figure 6. ADPKD in a 63-year-old man who presented with increasing upper abdominal pain and limited mobility ("inability to tie his shoes"). The patient had undergone left nephrectomy and kidney transplantation in 1987. (a) Coronal gradient-echo T1-weighted MR image shows typical low-signal-intensity fluid within multiple small cysts throughout the liver. High-signal-intensity fluid is seen within the dominant cyst (arrow), which involves the posterior right lobe superiorly. (b) Axial fast spin-echo T2-weighted MR image shows scattered intrahepatic cysts, all with similar high signal intensity. (c) Axial gradient-echo T1-weighted MR image obtained at the same level as b once again demonstrates high signal intensity of the dominant cyst (arrow), a finding that indicates possible complication from infection or hemorrhage. The patient was symptomatic but was a poor surgical candidate due to cardiac disease. On the basis of the imaging findings, temporizing percutaneous aspiration of the dominant cyst was selected as the best therapeutic option.







c.

At CT, MR imaging, and US, massive hepatomegaly with innumerable, predominantly simple cysts are

present. Morino et al (21) described several patterns of cyst distribution. Patients with PLD characterized by large cysts located primarily on the liver surface (type 1) respond favorably to extensive surgical cyst fenestration procedures, whereas those with innumerable small cysts located throughout the liver (type 2) generally obtain little symptomatic relief (21). Posterior segment cysts are also less amenable to fenestration, whether the procedure is open surgical (22) or laparoscopic (21), as are deeper cysts without overlying superficial cysts. MR imaging, US, and CT performed prior to fenestration with or without resection readily demonstrate cyst location and size and help identify islands of spared parenchyma (Fig 7) (23), which the surgeon should avoid when large numbers of cysts are to be fenestrated during a single procedure.

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Figure 7. Hepatic cysts in a 61-year-old man with symptomatic liver disease and laboratory test results indicating biliary obstruction. (a) Intravenous contrast–enhanced CT scan through the upper liver obtained in 2003 demonstrates complete replacement of the lateral segment of the left hepatic lobe by innumerable small cysts. (b) Contrast-enhanced CT scan through the upper liver obtained in 2005 demonstrates similar findings in the left hepatic lobe and new intrahepatic duct dilatation (arrow). Islands of normal parenchyma are seen in the right lobe. (c) Contrast-enhanced CT scan through the inferior right hepatic lobe demonstrates a very large cyst located along the surface. This dominant cyst appears amenable to fenestration. The small cysts in the lateral segment and the deep portion of the right lobe are not as amenable. On the basis of imaging findings, the patient underwent partial resection of the left lateral segment and anterior right lobe (nonsegmental resection plane), along with fenestration of the dominant cyst.





a.

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Vasculature

Portal vein patency should be assessed (Fig 8) (23). Compression of the main portal vein may result in portal hypertension (6) as well as associated findings such as splenomegaly and ascites (Fig 9). It may be difficult to identify the main portal vein and its branches owing to the distorted anatomy produced by the cysts. Identification of arterial variants is generally easier at intravenous contrast-enhanced CT or MR imaging, but careful attention to unenhanced CT scans may also sometimes reveal these findings for the transplantation surgeon.

Mass Effect on Adjacent Structures

Mass effect on the inferior vena cava (Fig 8), stomach (Fig 3), and diaphragm are well depicted with CT, US, and MR imaging, particularly with b.





multiplanar reformation or coronal imaging. MR imaging with MR cholangiopancreatography may demonstrate biliary dilatation associated with either peribiliary cysts or potentially treatable large intrahepatic cysts.



9a.

9b.

Figures 8, 9. (8) PLD and venous thrombosis. (a) Intravenous contrast–enhanced CT scan through the liver demonstrates marked intrahepatic compression of the inferior vena cava (arrow) in the caudate lobe. This compression resulted in stasis. (b) CT scan obtained inferior to a shows a nonocclusive thrombus (arrow) within the infrahepatic inferior vena cava. (c) Intravenous contrast–enhanced CT scan reveals thrombus (arrow) in the lateral segmental portal vein branches. (d) CT scan obtained at the hilum shows patent main and right portal veins without thrombus. The explanted liver weighed 4850 g and was 90% replaced by cysts. At surgery, multiple cysts had to be ruptured to mobilize the liver and remove it from the peritoneal cavity. The portal vein was elongated due to distortion by the cysts and had to be shortened to avoid kinking at the portal anastomosis. (9) Portal hypertension associated with PLD. (a) Longitudinal US image through the right hepatic lobe demonstrates multiple cysts of varying size and a moderate amount of ascites. (b) Longitudinal US image of the spleen demonstrates moderate splenomegaly.

Therapeutic Options

In addition to the chronic pain and immobility caused by massive hepatomegaly in patients with PLD, more acute complications may occur and require intervention. In one series in which 40 patients with polycystic livers were followed up over a nearly 5-year period, nine patients (22.5%) had cyst bleeding, five (12.5%) had cyst rupture, five (12.5%) had cyst infection, 12 (30%) required intervention, one (2.5%) developed portal hypertension, two (5%) received a liver transplant, and one (2.5%) died of complications from liver cysts (1). The patient who died was immunosuppressed from kidney transplantation for polycystic renal disease and died of gram-negative sepsis despite cyst aspiration and treatment with antibiotics (1).

Therapeutic options may focus on individual cyst therapy that provides temporizing relief of pain or mass effect; or on widespread cyst removal, which more frequently results in longterm relief of symptoms (Table). Temporizing therapies include cyst aspiration and sclerosis, cyst enucleation, or limited cyst fenestration. Worsening pain associated with rapid expansion of dominant infected or hemorrhagic cysts may respond well to this therapy. Patients may opt for needle aspiration while awaiting transplantation (24,25). Cyst enucleation (removal of the entire cyst and cyst wall) is an option but is generally reserved for patients with isolated hepatic cysts rather than PLD.

More permanent solutions for symptomatic PLD include widespread cyst fenestration, cyst fenestration with partial resection, and liver transplantation. Cyst fenestration, also referred to as unroofing or deroofing, may be performed laparoscopically or with an open surgical procedure. Fenestration involves wide excision of the protruding portion of the cyst wall, with the edge of the opened cyst sutured to adjacent structures to expose the inner surface to the peritoneum for drainage. Lin et al (26) first described open, wide fenestration of superficial cysts, allowing deeper cysts to be unroofed in a sequential, tunneled fashion during the same operation. Several groups have performed this extensive deroofing procedure laparoscopically, noting that the cysts should be punctured prior to fenestration to avoid injury to the often-stretched hepatic and portal veins (21,27). Conversion to laparotomy may be necessary secondary to adhesions from prior open cyst therapies (21). Over 30 cysts may be treated in a single procedure, and patients may lose over 2 L in cyst fluid. When treated with fenestration (unroofing), virtually all patients in reported series experienced immediate relief of symptoms (21, 27,28). However, the cysts will recur in 22%-75% of patients with PLD at an intermediate follow-up interval of 24 months (27,28), often with symptoms (2,15,21,27,28). Although the reported morbidity and mortality rates for open fenestration (0%-66% and 0%-11%, respectively) versus laparoscopic fenestration (29%-67% and 0%, respectively) are equivalent, symptomatic recurrence is higher for the laparoscopic technique (2). In general, fenestration is safe and acceptable for patients with a dominant cyst pattern in which liver size can be reduced after cysts collapse (29).

Cyst fenestration accompanied by partial hepatic resection (Fig 7) is a viable option for patients with a nonuniform distribution of cysts and some areas of spared parenchyma. Because of the distorted anatomy in these patients, segmental hepatic resection is technically challenging (22, 23). Although most patients experience persistent symptomatic relief at intermediate follow-up (mean, 20 months) (23), not all patients undergoing this combined technique experience immediate or complete pain relief (2,22). This phenomenon is probably a result of the type of liver involvement, which is more likely to be a pattern of diffuse and smaller cysts in these patients than in patients undergoing fenestration alone. The combination of resection-fenestration is suitable for patients with a heterogeneous cyst pattern; in this

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b.

c.

combined procedure, grossly affected segments with diffuse small cysts are resected in combination with fenestration of larger cysts in other areas of the liver to allow reduction in liver size (29).

Liver transplantation for PLD (Fig 10) was first reported by Starzl et al (9), who performed the procedure in patients with lethal exhaustion syndrome due to massive hepatomegaly. Because the evolution of liver disease in these patients is slow (22), most patients have lived with bilobar disease for many years prior to consideration for transplantation for PLD (9,16,24). Liver transplantation usually leads to excellent symptomatic relief in these patients but is a high-risk procedure, with associated mortality rates ranging from 10% to 33% (16). The high mortality rate reported in early PLD transplantation series may have been due to the severely debilitated and malnourished condition of these patients, who underwent liver transplantation only as a "last resort" (2). Nevertheless, according to published reports, **Figure 11.** Ascites and bile leaks after partial nonsegmental resection and cyst fenestration in the same patient as in Figure 7. (a) Intravenous contrast–enhanced CT scan obtained after removal of the majority of the lateral segment demonstrates postsurgical changes. A small amount of biliary air is present, and ascites is seen surrounding the spleen. (b) CT scan obtained inferior to a shows that portions of the anterior segment of the right hepatic lobe have been removed, and the dominant cyst has been opened. Soft tissue in the anterior aspect of the cyst (arrows) could represent redundant cyst wall or clot. (c) Spot radiograph obtained during endoscopic retrograde cholangiography demonstrates opacification of the intrahepatic ducts is seen near the surgical resection margin. (d) Magnified spot radiograph obtained during endoscopic retrograde cholangiography demonstrates extravasation of contrast material (arrow) from the left intrahepatic ducts in the anterior segment region near the surgical drain.



a.



the majority (50%-100%) of patients who undergo transplantation for PLD have normal hepatic function (9,16,30). Between 15% and 50% of patients have portal hypertension (9,30), and 25%-80% have undergone prior cyst aspiration or fenestration (3,9,16). Transplantation may be performed with cadaveric or living related donors,

although screening and informed consent regarding disease in the donor liver must be addressed (17). Liver transplantation has been performed with concurrent kidney transplantation in patients with ADPKD and renal failure who were also symptomatic from massive hepatomegaly. The cysts do not recur after transplantation in these patients due to the absence of von Meyenburg complexes in the donor liver.

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Figure 12. Massive hepatomegaly resulting in a redundant right hemidiaphragm. (a) Unenhanced CT scan through the midportion of the liver demonstrates innumerable cysts of varying size, with partial sparing of the periphery of segment IVa. The explanted liver was so large that the patient had to undergo reduction of the right hemidiaphragm at the time of transplantation because the donor liver kept falling into the chest during surgery. Arrow indicates the orientation of the celiac axis prior to transplantation. (b) Postoperative unenhanced CT scan through the midportion of the donor liver demonstrates a normal graft without complication. Splenomegaly is much more apparent now that the massively enlarged liver has been removed. Note the change in orientation of the celiac axis (arrow) after removal of the polycystic liver.







Complications

Abdominal CT with (creatinine-permitting) or without intravenous contrast material is generally used to assess complications of the various cyst therapies. Massive perioperative fluid losses during extensive cyst removal may lead to severe dehydration (6). Complications of cyst fenestration include ascites in all patients (6,22,26), generally diminished by the third day after surgery when a drain is left in place (27) or 5-26 days after surgery when diuretics are used (23). Persistent ascites after fenestration is due to the inability of the peritoneum to absorb more than 900 mL of fluid per day (6). If trocar insertion sites are not carefully closed in patients undergoing laparoscopic procedures, infiltration of ascites into the abdominal wall may occur (21). Pleural fluid accumulation occasionally requires thoracentesis (21,23, 27). Bile leak (Fig 11), hemorrhage, and extravasation of ascites into the subcutaneous tissues may also be seen (27) following extensive fenestration of cysts. Imaging at intermediate followup (24 months) may show recurrence of (often smaller) cysts at the site of previous treatment (15,23) or new dominant or symptomatic cysts (15,27) anywhere in the liver. Follow-up CT at this interval generally reveals hypertrophy of previously spared hepatic regions (23).

Liver transplantation in these patients is associated with the usual intra- and postoperative complications. Difficulties related to the massive size of the explanted liver, including extensive cyst puncture to mobilize the liver and remove it from the peritoneal cavity, may also be encountered (Figs 8, 10, 12). Transplantation in PLD patients who have undergone fenestration or resection may be hindered by adhesions from these earlier procedures (2). Combined liver-kidney transplantation presents greater technical difficulties for the surgeon (31), and these patients are at greater risk for hemorrhage (16,31) and death (31).

Conclusions

PLD is characterized by progressive development of fluid-filled biliary epithelial cysts throughout all segments of the liver. Not all patients with polycystic livers have polycystic renal disease, although the majority do. Massive hepatomegaly may lead to incapacitating symptoms, ultimately requiring liver transplantation. Cyst size and patterns of cyst distribution may affect the therapeutic options used in these patients prior to transplantation.

Teaching Point

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In most reported series, the majority of patients treated for PLD have ADPKD.

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However, symptomatic polycystic liver patients have massive hepatomegaly.

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Cyst size and patterns of cyst distribution affect therapeutic options.

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Therapeutic options may focus on individual cyst therapy that provides temporizing relief of pain or mass effect; or on widespread cyst removal, which more frequently results in long-term relief of symptoms (Table).

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Abdominal CT with (creatinine-permitting) or without intravenous contrast material is generally used to assess complications of the various cyst therapies.