Management of Nonparasitic Hepatic Cysts

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BACKGROUND: The optimal management of nonparasitic hepatic cysts (NPHC) is a topic of debate. The purpose of this study was to evaluate our 17-year experience with NPHC.

STUDY DESIGN: From January 1990 to August 2007, 131 consecutive patients with NPHC were evaluated and treated at our institution. Seventy-eight patients (60%) had simple hepatic cysts (SHC). The remaining 53 (40%) had polycystic liver disease (PLD). Morbidity, mortality, and recurrence rates for each of the two groups were evaluated.

RESULTS: Thirty-seven patients underwent open deroofing (SHC, 24; PLD, 13), 66 had laparoscopic deroofing (SHC, 46; PLD, 20), 19 had percutaneous drainage (SHC, 4; PLD, 15), 3 had major hepatic resections (PLD, 3), 4 had cystojejunostomy (SHC, 4), and 2 had combined hepatorenal transplantation (PLD, 2). Corresponding morbidity, mortality, and recurrence rates were, respectively: conventional deroofing: SHC, 29%, 0%, 8%; PLD, 8%, 0%, 0%; laparoscopic deroofing: SHC, 2%, 0%, 2%; PLD, 25%, 0%, 5%; percutaneous drainage: SHC, 0%, 0%, 75%; PLD, 0%, 0%, 20%; cystojejunostomy: SHC, 75%, 0%, 25%; major hepatic resections: PLD, 66%, 0%, 0%; and hepatorenal transplantation: PLD, 50%, 50%, 0%.

CONCLUSIONS: Laparoscopic deroofing provided complete relief of symptoms for both SHC and PLD. Percutaneous drainage was our procedure of choice for infected liver cysts and potentially for patients who cannot tolerate general anesthesia. Liver and liver-kidney transplantations were reserved for patients with end-stage PLD alone and in association with end-stage renal disease, respectively. (J Am Coll Surg 2009;209:733–739. © 2009 by the American College of Surgeons)

Nonparasitic hepatic cysts (NPHC) present with a broad range of anatomic and clinical manifestations. Although laparoscopic and percutaneous drainage constitute minimally invasive therapeutic options, resection, fenestration, and liver transplantation allow for the treatment of massive liver enlargement. Differentiation between simple hepatic cysts (SHC) and adult polycystic liver disease (PLD) is essential to determine the optimal therapeutic approach.¹ The purpose of this study was to evaluate our 17-year experience treating NPHCs.

METHODS

Between January 1990 and August 2007, 131 consecutive patients with NPHCs were evaluated and treated at our institution. There were 41 men and 90 women, with a mean age of 62.5 years (range 19 to 87 years). All patients underwent an initial ultrasonography diagnostic screening. The surgical approach (deroofing or resection) was based on the anatomic information provided by CT scans. Polycystic liver lesions complicated by bleeding or infection were evaluated by additional ultrasonography or MRI examinations. Malignancy was suspected in cases of cystic wall thickening or papillary growth within the cyst. Contrast-enhanced CT scans combined with MRI and α-fetoprotein, CEA, and CA 19-9 serum levels were used for preoperative diagnoses. Oncologic resections were based on intraoperative findings. Malignant cystic tumors were not included in this study.

Patients were divided into two groups according to cyst type: simple hepatic cysts and polycystic liver disease. Table 1 depicts their demographic information. Symptoms at presentation, therapeutic procedures, postoperative complications according to Dindo’s classification,² and short-
and longterm results were considered when performing our analysis of prospectively collected data.

### Simple hepatic cysts

Simple hepatic cysts, with a mean diameter of 9.2 cm (range 3 to 20 cm), were encountered in 78 patients. Forty-three cases involved the right lobe and 29 the left lobe. Six were bilateral. Presenting symptoms are summarized in Table 1. In 20 instances (25.6%), initial findings were associated with cyst complications: 11 bleeds, 8 infections, and 1 spontaneous rupture. Therapeutic procedures are shown in Table 2. Elective surgical deroofing, as described in an open fashion by Lin and colleagues and laparoscopically by Vauthey and associates, was the procedure of choice. Only the cyst wall (rather than compressed liver parenchyma) was resected to avoid postoperative bile leaks. Determination of an open or laparoscopic approach was based solely on the experience of the surgeon. In open cases, a right subcostal incision was made, intraoperative ultrasonography was not routinely performed, and resection of the cystic wall was carried out with electrocautery.

Intraoperative cholangiograms were done only when the cystic contents suggested a biliary communication. Abdominal drains were always used after deroofing. Resected cystic wall specimens were routinely sent for pathologic evaluation to exclude malignancies. In laparoscopic cases, a 10- to 12-mm port was inserted at the umbilicus, and additional ports were positioned according to the location of the cyst. In most instances, a second port was placed in the epigastric area, and a third one in the right or left upper quadrants. During our early laparoscopic experience, the cyst wall was resected with a monopolar hook and scissors. Most recently, we have opted for an ultrasonic scalpel. Four patients who presented with infected cysts underwent percutaneous drainage with multipurpose 8- to 12-F drains. Roux-en-Y cystojejunostomies were performed in four cases of giant cysts located in the posterior aspect of the right lobe of the liver. Three of these four patients had recurrent cysts after unsuccessful open deroofing attempts (two of them at outside hospitals).

### Polycystic liver disease

There were 53 patients with PLD who were divided into three groups according to Gigot’s classification. Group I (n = 33) included patients with a few large cysts. Group II (n = 14) was composed of patients with diffuse cystic involvement that retained areas of preserved hepatic parenchyma. Group III (n = 6) encompassed patients with massive cystic involvement and little uninvolved parenchyma. Cysts had a mean diameter of 11.5 cm (range 3 to 25 cm) and were predominant in the right lobe in 9 patients, in the left lobe in 3, and bilateral in 41. Presenting symptoms are detailed in Table 1. In nine patients (20.7%), initial findings were associated with cyst complications: four bleeds and five infections. Treatment modalities (Table 2) were based on evaluation of both clinical symptoms and Gigot’s classification. Open and laparoscopic surgical deroofing was performed according to the techniques described in the preceding text. Patients selected for this procedure were mostly in groups I and II. Only dominant and symptomatic lesions were deroofed. Percutaneous drainage was reserved for infected cysts (as identified by ultrasonography or MRI or both) and for patients who could not tolerate general anesthesia. Multipurpose drains were used in all cases. Patients with massive hepatomegaly associated with respiratory and digestive impairment underwent liver resections through a bilateral subcostal incision with a midline extension. Liver transection was with an ultrasonic

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**Table 1. Demographic and Clinical Characteristics**

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Simple cysts</th>
<th>Polycystic liver disease</th>
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<tbody>
<tr>
<td>n</td>
<td>78</td>
<td>53</td>
</tr>
<tr>
<td>Age, y (range)</td>
<td>61.3 (29–87)</td>
<td>64 (22–85)</td>
</tr>
<tr>
<td>Gender, male:female</td>
<td>19:59</td>
<td>14:39</td>
</tr>
<tr>
<td>Pain, n (%)</td>
<td>45 (57.7)</td>
<td>32 (60.3)</td>
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<tr>
<td>Nausea/vomiting/early satiety, n (%)</td>
<td>12 (15.4)</td>
<td>3 (5.6)</td>
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<tr>
<td>Fatigue/dyspnea, n (%)</td>
<td>9 (11.5)</td>
<td>7 (13.2)</td>
</tr>
<tr>
<td>Abdominal mass, n (%)</td>
<td>13 (16.6)</td>
<td>6 (11.3)</td>
</tr>
<tr>
<td>Fever, n (%)</td>
<td>5 (6.4)</td>
<td>5 (7.54)</td>
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<tr>
<td>Diameter of dominant cyst, cm (range)</td>
<td>9.2 (3–20)</td>
<td>11.5 (3–25)</td>
</tr>
</tbody>
</table>

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**Table 2. Therapeutic Procedures in Patients with Simple Hepatic Cysts and with Polycystic Liver Disease**

<table>
<thead>
<tr>
<th>Therapeutic procedures</th>
<th>Patients with simple hepatic cysts (n = 78)</th>
<th>Patients with polycystic liver disease (n = 53)</th>
</tr>
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<tr>
<td>n</td>
<td>%</td>
<td>n</td>
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<tr>
<td>Open deroofing</td>
<td>24</td>
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<tr>
<td>Laparoscopic deroofing</td>
<td>46</td>
<td>58.9</td>
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<tr>
<td>Percutaneous drainage</td>
<td>4</td>
<td>5.12</td>
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<tr>
<td>Cystojunostomy</td>
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<td>5.12</td>
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<tr>
<td>Hepatectomy</td>
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<td>5.7</td>
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<tr>
<td>Hepatorenal transplant</td>
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<td>3.8</td>
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dissector. We performed a left hepatectomy, a right trisectionectomy, and one nonanatomic resection. Two patients with type III disease who presented with massive hepato-megaly and end-stage renal failure underwent combined liver-kidney transplantation. One of them had previously undergone bilateral nephrectomies.

RESULTS

Simple hepatic cysts

Conventional deroofing
Mean hospital stay was 7.4 days (range 2 to 19 days). There were seven short-term postoperative complications: one intra-abdominal abscess successfully treated by percutaneous drainage, one bile leak that resolved spontaneously after percutaneous drainage, four cases of small bowel obstruction treated nonsurgically with nasogastric decompression, and one pneumothorax (5 grade II according to Dindo’s classification, 2 grade IIIa). Cyst recurrences developed in two patients (5.2%) after 21 and 35 months. A third patient presented with an incisional hernia.

Percutaneous drainage
All four patients who underwent percutaneous drainage presented with cysts originating from infected cysts, and required antibiotics tailored to the organisms identified by culture (Escherichia coli in three patients and Klebsiella in the remaining one). All patients responded favorably to treatment. The percutaneous drains were removed once both clinical and radiologic resolution was achieved. Recurrent lesions developed in three patients (75%). One asymptomatic patient required no treatment. Another underwent laparoscopic deroofing. The third was treated with a cystojejunostomy. Except for the recurrences, there was no associated morbidity or mortality. Mean hospital stay was 7 days (range 2 to 15 days).

Cystojejunostomy
Four patients with giant lesions located in the right posterior lobe of the liver underwent conventional cystojejunostomies. FEVERS developed in three suspected to originate from bacterial contamination of the remaining cavity and were successfully treated with empiric oral antibiotics. In one case, a persistent symptomatic lesion required additional percutaneous drainage and alcoholization. There were no recurrences. Mean hospital stay was 9.2 days (range 3 to 29 days).

Mean followup for the simple hepatic cysts group was 43 months (range 11 to 98 months). There was no mortality, and the incidence of morbidity was 8.9%. Mean hospital stay for laparoscopic deroofing was significantly lower than that for conventional deroofing (p ≤ 0.05). There were no differences in morbidity or mortality between laparoscopic and open approaches.

Polycystic liver disease

Open deroofing
Open deroofing was performed in 13 patients with Gigot’s type I and II disease. A patient with type III PLD who underwent open deroofing of multiple cysts during our early experience, experienced ascites and renal and respiratory failure unresponsive to medical treatment. An emergency combined liver-kidney transplantation was undertaken, but the patient died postoperatively from a cardiac arrest. Mean hospital stay was 8.6 days (range 7 to 17 days).

Laparoscopic deroofing
Laparoscopic deroofing was performed in 20 patients. Only large dominant and symptomatic cysts were treated, as with the open approach. One patient (5%) with type II Gigot’s disease experienced a recurrence and was treated with a second laparoscopic fenestration. She eventually underwent bilateral nephrectomies and a combined liver-kidney transplantation. Mean hospital stay was 3 days (range 1 to 6 days). The incidence of morbidity for deroofing among PLD patients was 9.4%. There was 1 case each (n = 5) of postoperative bleed, small bowel obstruction, acute renal failure, post drainage bacteremia, and soft tissue infection. All were grade II. The mean hospital stay for laparoscopic deroofing was significantly lower than that for conventional deroofing (p ≤ 0.05). There were no differences in morbidity between the open and laparoscopic approaches.

Percutaneous drainage
Fifteen patients presented with infected cysts and systemic sepsis. All underwent successful treatment with percutaneous drainage and received intravenous antibiotics. Culture results showed gram-negative bacilli (E coli, Klebsiella, Enterobacter, and Serratia) in all instances. Drains were removed upon resolution of clinical symptoms and radiologic findings. There were three (20%) symptomatic recurrences. One was treated with laparoscopic deroofing. The remaining two were drained percutaneously for a second time, in one occasion with additional alcohol sclerosis. There was no morbidity or mortality.
**Liver resection**

Liver resection was the treatment of choice in three selected patients with massive hepatomegaly and type III PLD who presented with gross abdominal distension and weight loss. Procedures performed included a left hepatectomy, a right trisectionectomy, and a nonanatomic resection. One of the three is currently listed for a combined liver-kidney transplantation because of continued hepatic enlargement and deterioration of renal function. There were two complications. One patient required reexploration for bleeding and hemodialysis for acute renal failure (grade IVa). Persistent ascites developed in another requiring prolonged hospitalization (grade II). Mean hospital stay was 17.3 days (range 5 to 28 days).

**Combined liver-kidney transplantation**

Combined liver-kidney transplantation was undertaken in two patients with PLD. One was transplanted emergently after experiencing severe ascites after deroofing of a dominant cyst. He died post transplantation of a cardiac arrest (grade V). The second had previously undergone two laparoscopic deroofings and bilateral nephrectomies. She is currently 3 years posttransplantation, alive and well with normal liver and kidney function.

Mean followup time for the entire PLD group was 59.5 months (range 17 to 96 months).

**DISCUSSION**

The differential diagnosis between parasitic cysts, pseudocysts, true cysts, and malignant cysts may be difficult.1 Echinococcal (hydatid) cysts represent the major pathology to exclude in many regions of the world, such as in Argentina. Although epidemiologic data, serologic results, and imaging studies are frequently useful, when in doubt we perform (like many other authors) a laparoscopic exploration. If a diagnosis of hydatidosis is made,6 we convert to an open approach for resection. Pseudocysts (intrahepatic bilomas and abscesses) are infrequent lesions, devoid of epithelial lining, that often develop as complications of hepatic surgery and abdominal trauma.7 Simple hepatic cysts originate in embryologic blind ducts with no biliary tree communication. They have an epithelial cover with serous contents, and can be divided into two different types: simple cysts and PLD.4 Simple cysts can be single or multiple. PLD has a familial component, and may involve other organs in addition to the liver. Neither carries malignant potential. Symptoms develop as a result of mass effect or secondary to complications such as bleeding and infections.8 Treatment is reserved for symptomatic or complicated cysts, or when malignancy cannot be excluded.9

Multiple reports confirm the effectiveness of surgical deroofing for hepatic cysts. This technique, originally described by Lin and coworkers3 in 1968, entails the resection of the largest possible amount of emergent cystic wall. Laparoscopic deroofing, with lower morbidity rates, similar recurrence rates, and shorter hospital stays than its open counterpart,10-12 has become the treatment of choice for both symptomatic large simple and PLD lesions.13,14 Patients treated with the conventional open technique in our series, as in many other institutions, represent the laparoscopic learning curve. When deroofing, special care should be taken not to resect hepatic parenchyma, given that transected bile ducts may lead to postoperative bile leaks (Fig. 1). In their impressive series, Gamblin and colleagues15 used vascular staplers or tissue sealing devices and considered a hand-assisted technique for large lesions requiring extensive liver mobilization. In our work, we opted for monopo-
lar scissors or ultrasonic scalpel to minimize our costs without compromising patient safety. Some authors recommend filling the cyst cavity with omentum to achieve a better drainage of the epithelial secretions into the peritoneal cavity. We do not routinely perform this maneuver. We believe that our series provides additional support to the previous observations by Gamblin and coworkers that laparoscopic resections have become the standard of care for the treatment of benign symptomatic cysts.

Percutaneous drainage of symptomatic hepatic cysts has been popularized recently. Because recurrence after simple puncture and aspiration of the cyst is almost guaranteed, the use of sclerosing substances after evacuation of the cyst has been advocated as a way to improve recurrence rates. Such substances, however, are formally contraindicated in cases of suspected or proven cyst-biliary tree communications or in infected cysts. We use this approach selectively, mostly for infected lesions, in patients with systemic manifestations. In all cases, we obtained prompt relief of septic conditions.

Large cysts in the right posterior lobe have a high recurrence rate. In these lesions, the intimate contact between liver and diaphragm prevents an adequate continuous drainage of the deroofed cyst into the peritoneal cavity, leading to reaccumulation of its contents. Three of the four posterior right giant cysts that we treated with Roux-en-Y limbs had had previous unsuccessful deroofing attempts. Three resolved promptly, as documented by postoperative CT scans. The fourth patient presented with a symptomatic persistent lesion 1 month later and was successfully treated with additional percutaneous drainage and alcoholization. Three of these patients presented fever during the postoperative period, and they were all successfully treated with empiric oral antibiotics. We believe that this unusual treatment may prove helpful to prevent recurrence of large posterior cyst. Our experience also seems to suggest that antibiotics should be routinely administered postoperatively after this procedure.

PLD exhibits major differences. It is characterized by frequent renal involvement and an autosomal dominant familial history with high genetic penetrance. PLD cysts are multiple and tend to increase in number and size throughout life, as opposed to simple cysts. Because deterioration of hepatic function is very rare, prognosis is usually determined by the concomitant renal disease. Cyst infection, a life-threatening condition in patients with PLD, usually presents with severe systemic deterioration superimposed on chronic renal failure. We successfully treated all these patients with prompt percutaneous drainage of the infectious focus, minimizing any surgical aggression. Correct identification of the infected cysts may be difficult. CT scanning usually provides a view of the anatomy, but MRI or ultrasonography examination at the time of drainage may be necessary to identify their heterogeneous content (Fig. 2). The advantage of MRI in these cases is based on the fact

![Figure 2. Complex cyst in a patient with polycystic liver disease as viewed by (A) CT scan and (B) ultrasound imaging. Heterogeneous contents (arrows) in one of the cysts can be clearly identified by ultrasound.](image)
that the high protein content of complicated cysts confers on them a characteristic hyperintensity in T1-weighted sequences.7

Symptoms in PLD type I are usually from the mass effect of a few well-defined lesions. Laparoscopic deroofing has been effective in our experience and in that of other authors,5,14,25,28 and should be considered as the first choice approach for this group of patients.29 Laparoscopy is associated with fewer adhesions, facilitating reoperations should more interventions be necessary.30

Finally, there is a third group of patients, Gigot’s PLD types II and III, with symptomatic massive hepatomegaly. Deroofing does not offer great relief to the respiratory failure, digestive symptoms, and cachexia of this group.5,25 Our first patient experienced severe respiratory and renal insufficiency soon after the procedure, prompting an emergency hepatorenal transplantation. Combined hepatic resection and cyst fenestration in the remnant liver seems to be a more appropriate option in these patients.27,31,32 Symptomatic relief with low morbidity was achieved in most cases.31,33,34 We undertook this approach in three patients, with good results.

In patients with type III disease showing diffuse liver enlargement and small cysts not suitable to deroofing or resection, direct evaluation for liver transplantation should be considered.26,35 Nevertheless, liver transplantation continues to have a limited role in the management of this disease. In spite of many series reporting good results, PLD does not usually evolve into end-stage liver disease, making this decision difficult. Transplantation must be balanced against the risks of operation, immunosuppression, quality of life, and possible benefits of a combined renal implant.36–38 We performed two combined transplantations. One patient who received a transplant emergently died as a result of a cardiac arrest. The other remains alive and well with no complications.

Various liver malignancies may present as cystic lesions. During the time period of our study, we diagnosed seven patients with cystic neoplasms. All of them underwent major hepatic resections. Final pathologic diagnoses were: two cystoadenocarcinomas, two cystic sarcomas, one cystic hepatocarcinoma, one hepatic adenoma with areas of severe dysplasia in the setting of Caroli’s disease, and one cystic adenoma. Tumor cavities generally developed as a result of necrosis and subsequent liquefaction of the malignant mass, induced by rapid cellular proliferation or local and systemic chemotherapy. The presence of thickened or irregular cyst walls should raise the suspicion of a neoplasm. When in doubt, intraoperative ultrasonography and frozen examination of surgical specimens is recommended. If a malignant diagnosis is confirmed, complete resection of the lesion with oncologic margins is required whenever possible. Outcomes depend on the type of tumor present.

Laparoscopic deroofing provided complete relief of symptoms for both SHC and PLD. Percutaneous drainage was our procedure of choice for infected liver cysts and potentially for patients who could not tolerate general anesthesia. Liver and liver-kidney transplantation was reserved for cases of end-stage PLD alone and in association with end-stage renal disease, respectively.

**Author Contributions**

Study conception and design: Mazza, Fernandez, Pekolj, de Santibañes

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Analysis and interpretation of data: Mazza, Fernandez, Pekolj, de Santibañes

Drafting of manuscript: Mazza, Fernandez, Molmenti

Critical revision: Molmenti, de Santibañes

**REFERENCES**


