A 20-Year Experience with Liver Transplantation for Polycystic Liver Disease: Does Previous Palliative Surgical Intervention Affect Outcomes?

John T Baber, MD, Jonathan R Hiatt, MD, FACS, Ronald W Busuttil, MD, PhD, FACS, Vatche G Agopian, MD

BACKGROUND: Although it is the only curative treatment for polycystic liver disease (PLD), orthotopic liver transplantation (OLT) has been reserved for severely symptomatic, malnourished, or refractory patients who are not candidates for palliative disease-directed interventions (DDI). Data on the effect of previous DDIs on post-transplant morbidity and mortality are scarce. We analyzed the outcomes after OLT for PLD recipients, and determined the effects of previous palliative surgical intervention on post-transplantation morbidity and mortality.

STUDY DESIGN: We performed a retrospective analysis of factors affecting perioperative outcomes after OLT for PLD between 1992 and 2013, including comparisons of recipients with previous major open DDIs (Open DDI, n = 12) with recipients with minimally invasive or no previous DDIs (minimal DDI, n = 16).

RESULTS: Over the 20-year period, 28 recipients underwent OLT for PLD, with overall 30-day, 1-, and 5-year graft and patient survivals of 96%, 89%, 75%, and 96%, 93%, 79%, respectively. Compared with the minimal DDI group, open DDI recipients accounted for all 5 deaths, had inferior 90-day and 1- and 5-year survivals (83%, 83%, and 48% vs 100%, 100%, 100%; p = 0.009), and greater intraoperative (42% vs 0%; p = 0.003), total (58% vs 19%; p = 0.031), and Clavien grade IV or greater (50% vs 6%; p = 0.007) postoperative complications, more unplanned reoperations (50% vs 13%; p = 0.003), and longer total hospital (27 days vs 17 days; p = 0.035) and ICU (10 days vs 4 days; p = 0.045) stays.

CONCLUSIONS: In one of the largest single-institution experiences of OLT for PLD, we report excellent long-term graft and patient survival. Previous open DDIs are associated with increased risks of perioperative morbidity and mortality. Improved identification of PLD patients bound for OLT may mitigate perioperative complications and potentially improve post-transplantation outcomes. (J Am Coll Surg 2014;219:695–703. © 2014 by the American College of Surgeons)

Polycystic liver disease (PLD) is a rare but debilitating autosomal dominant disorder in which cysts of varying size form throughout the liver. This disorder can occur in isolation (autosomal dominant polycystic liver disease), or more commonly, with renal cysts (autosomal dominant polycystic kidney disease).[1] Although typically asymptomatic, a small fraction of PLD patients will develop compressive symptoms due to massive hepatomegaly, resulting in abdominal or back pain, dyspnea, or early satiety. Severely affected patients can develop malnutrition, ascites, and lower extremity edema secondary to compression of the hepatic veins, portal vein, or inferior vena cava, and jaundice secondary to extrahepatic bile duct compression.[2,3]

Orthotopic liver transplantation (OLT), the only curative treatment for PLD,[4-8] has been reserved for severely symptomatic patients with malnutrition and cachexia that preclude or are refractory to nontransplant interventions. These "disease-directed interventions" (DDIs) include percutaneous or laparoscopic cyst aspiration, open or laparoscopic cyst fenestration, and hepatic resection.[9-14] Citing concerns regarding donor organ scarcity and the risks of

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From the Departments of Surgery, Division of General Surgery (Baber, Hiatt), and Dumont-UCLA Transplant and Liver Cancer Centers, Pfleger Liver Institute (Busuttil, Agopian), David Geffen School of Medicine at the University of California, Los Angeles, Los Angeles, CA.

Correspondence address: Vatche G Agopian, MD, Division of Liver and Pancreas Transplantation, Department of Surgery, David Geffen School of Medicine at UCLA, University of California, Los Angeles, 10833 Le Conte Ave, Room 77-123 CHS, Box 957054, Los Angeles, CA 90095-7054. email: vagopian@mednet.ucla.edu

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Abbreviations and Acronyms

DDI = disease-directed intervention
HAT = hepatic artery thrombosis
MELD = Model for End-Stage Liver Disease
OLT = orthotopic liver transplantation
PLD = polycystic liver disease
SLK = simultaneous liver-kidney transplantation

lifelong immunosuppression, some authors have argued for aggressive surgical intervention as an alternative to OLT in all but the most severe cases.[4,13,15] Although effective in palliating symptoms, such an approach is associated with significant postoperative morbidity and may complicate future liver transplantation.[5,6,16-20]

Based on these previous observations, we hypothesized that recipients with previous open DDIs who required OLT for refractory PLD symptoms would be at greater risk for perioperative morbidity and mortality compared with PLD recipients who had not had major previous DDIs. Hence, the specific aims of this study were to analyze our experience with OLT for PLD and determine the effects of previous palliative surgical intervention on the outcomes after OLT.

METHODS

Using a prospectively collected database, we performed a retrospective review of all patients who underwent OLT for PLD at the University of California, Los Angeles (UCLA) between October 29, 1992 and June 25, 2013. The mean duration of post-transplant follow-up was 5.3 years. The study was approved by the UCLA Institutional Review Board.

Polycystic liver disease was diagnosed based on patient history and abdominal imaging demonstrating multiple simple cysts (>20) within the liver parenchyma, without

Table 1. Pretransplantation Characteristics

Variables	All patients (n = 28)	Open DDI (n = 12)	Minimal DDI (n = 16)	p Value*
Age, y†	54	55	52	0.798
Female, %	82	75	88	0.412
Isolated liver cysts, %	7	8	6	0.840
Concomitant renal cysts, %	93	91	94	0.840
Gigot class, median	3	3	3	NS
Symptoms, %				
Abdominal pain	93	83	100	0.097
Early satiety	57	46	69	0.242
Dyspnea	36	58	19	0.031
Ascites	29	25	31	0.729
Fatigue	18	33	6	0.068
Malnutrition	18	8	25	0.271
Portal hypertension	14	17	13	0.766
Liver failure	14	25	6	0.173
Comorbidities, %				
Diabetes	4	0	6	0.999
Hypertension	64	67	63	0.823
Hyperlipidemia	11	8	13	0.736
Smoking	43	50	38	0.523
Laboratory studies				
Total bilirubin, mg/dL†	0.6	0.8	0.5	0.031
International normalized ratio†	1.1	1.2	1.1	0.158
Serum creatinine, mg/dL†	1.5	1.8	1.5	0.399
Calculated MELD score†	10	15	11	0.245
Albumin, mg/dL	3.6	3.6	3.6	0.804
Pretransplant dialysis, %	21	25	19	0.703
Mechanical ventilation, %	4	8	0	0.256
Vasopressors, %	0	0	0	NS

*Indicates comparison of open DDI and minimal DDI groups.
†Non-normally distributed continuous variables, median values reported.
DDI, disease-directed intervention; MELD, Model for End-Stage Liver Disease; NS, not significant.

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