# Outcomes and Durability of Hepatic Reduction after Combined Partial Hepatectomy and Cyst Fenestration for Massive Polycystic Liver Disease

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**BACKGROUND:** Partial hepatectomy and cyst fenestration (PHCF) selectively provides clinical benefit in highly symptomatic patients with polycystic liver disease (PLD). This study aims to ascertain whether the reduction in liver volume (LV) achieved by PHCF is sustained long term.

**STUDY DESIGN:** Clinical data were retrieved from the electronic records of all patients with PLD who underwent PHCF between 1985 and 2014. Preoperative LVs (LV1), postoperative LVs (LV2), and late follow-up LVs (LV3) were measured from magnetic resonance or CT images.

**RESULTS:** Among 186 patients who underwent PHCF, 91% were Caucasian women with autosomal dominant polycystic kidney disease with a mean age of 49 years. Major perioperative complications (Clavien III/IV) occurred in 21% of the patients. Operative mortality (<90 days) was 2.7%. Eleven patients had liver failure develop, received liver transplants, or had liver-related deaths. Overall survival was 95.7%, 93.3%, 85.6%, and 77.7% at 1, 5, 10, and 15 years respectively. Imaging records for volumetry were unavailable in 32 patients. Of the remaining 154 patients, 34 had imaging for 1 LV, 64 for 2 LVs, and 55 for all 3 LVs. Median LV was 6,781 mL (interquartile range 4,903 to 8,341 mL) preoperatively and 2,502 mL (interquartile range 2,089 to 3,136 mL) after PHCF, leading to a median postoperative LV reduction of 61%. At follow-up (mean 8 years), median LV was 2,519 mL (interquartile range 2,083 to 3,752 mL). Interestingly, 33 of 62 patients with available LV2 and LV3 showed additional regression in LV at follow-up (median −14.1%), and the rest showed mild growth of 9.9%. Overall volumetric comparison of preoperative with follow-up liver imaging showed sustained LV reduction (median 61%).

**CONCLUSIONS:** Sustained long-term reductions in LV after PHCF can be achieved in selected patients with severe, highly symptomatic PLD. In our experience, liver-related death and subsequent liver transplantation are infrequent after PHCF. (J Am Coll Surg 2016;223:118−128. © 2016 by the American College of Surgeons. Published by Elsevier Inc. All rights reserved.)

Polycystic liver disease (PLD) is characterized by the presence of multiple cholangiocyte-derived epithelial cysts that cause progressive liver enlargement. Most commonly, PLD co-exists with autosomal dominant polycystic kidney disease (ADPKD),[1] and occurs less commonly as a genetically distinct disease with few or no renal cysts in autosomal dominant polycystic liver disease.[2] Polycystic liver disease is one of the most common extrarenal manifestations of ADPKD[3,4] and is defined clinically by the presence of any liver cyst. Hepatic cyst prevalence and total hepatic cyst volume increase with age and in women compared with men. Hepatic cysts are evident on MRI in 94% of patients with ADPKD who are older than 35

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**Abbreviations and Acronyms**

ADPKD = autosomal dominant polycystic kidney disease
IQR = interquartile range
LV = liver volume
LV1 = preoperative liver volume
LV2 = postoperative liver volume
LV3 = liver volume at follow-up
MELD = Model for End-Stage Liver Disease
PHCF = partial hepatectomy and cyst fenestration
PLD = polycystic liver disease

years of age.[1,5] Although most patients are asymptomatic initially, extensive PLD can lead to altered hepatic-related biochemical features and affect quality of life.[5] Cystic enlargement causes liver enlargement, which can be marked and result in dyspnea, early satiety, gastroesophageal reflux, mechanical back pain, hepatic venous outflow obstruction, portal vein and inferior vena cava compression, and, rarely, jaundice from bile duct compression.[6] When lifestyle is impaired substantially, surgical intervention is indicated to alleviate symptoms and restore quality of life. Partial hepatectomy and cyst fenestration (PHCF) selectively provides clinical benefit in highly symptomatic patients with massive hepatomegaly with an acceptable surgical risk when performed by an experienced liver surgeon. Our center has reported short-term and long-term effectiveness of PHCF for symptom control on limited number of patients previously.[7,8] This study aims to ascertain whether the reduction in liver volume (LV) achieved by PHCF is sustained long-term and to assess operative risk and survival.

## METHODS

Between July 1985 and April 2014, 186 patients with PLD underwent PHCF at Mayo Clinic Rochester, MN. Demographic characteristics and clinical data were reviewed retrospectively. Hepatic resection was offered to patients who had massive and symptomatic PLD that resulted in decreased clinical performance status, quality of life, and, in some cases, complications such as cholestasis and hepatic venous outflow obstruction. The primary indication for PHCF was the patients' decision that their lifestyle impairment from liver enlargement precluded or severely limited physical activity, social interaction, employment, or combinations of these findings. Partial hepatectomy and cyst fenestration was undertaken if at least one hepatic section (sector) was relatively spared of PLD in comparison with those sections diffusely involved with PLD, afferent and efferent hepatic vasculature was patent, and hepatic function was maintained. Preoperative evaluation of patients for resection has been detailed previously and routinely included hepatic, renal, and cerebrovascular imaging.[8] In brief, patients undergoing PHCF had type C PLD based on our earlier classification scheme.[8] Patients with PLD who underwent hepatic operations for reasons other than control of volume-related symptoms with or without concurrent complications from liver cysts were specifically excluded. All patients were operated on by a single surgeon (DMN). Perioperative morbidity included complications within the hospital stay or within 30 days of operation. Early mortality was defined as death within the hospital stay or within 90 days of operation. Clinical follow-up was defined as last recorded communication or visit with the patient. Imaging follow-up was defined as the last imaging study available at long-term follow-up (>1 year). Ten patients were enrolled into clinical trials of somatostatin analogues for severe PLD, 4 several years preoperatively (the administration of the somatostatin analogue was discontinued at the time of the surgery) and 6 postoperatively (postoperative liver images included in these patients were obtained before the initiation of somatostatin analogue therapy). Survival status was obtained on all patients using vital records website (www.archives.com). Patient survival was analyzed using the Kaplan-Meier method. Cause of death was obtained from medical records or through obtaining the official death certificate. The study was approved by the Mayo Clinic Rochester IRB and research authorization was provided by all patients. Radiographic volumetric measurements of the liver were performed on axial or coronal CT or MRI studies using Analyze software (Analyze 120.0, Biomedical Image Resource, Mayo Clinic). Preoperative LVs (LV1), postoperative LVs (<6 months, LV2), and late follow-up LVs (>1 year, LV3) were measured when available. Data are reported as mean ± SD for normally distributed data or median and interquartile range (IQR) for skewed data. Comparisons between groups were done by Student's *t*-test (2 groups) or ANOVA (more than 2 groups) for normally distributed data and by Wilcoxon rank sum test or Kruskal-Wallis test as appropriate for skewed data.

## RESULTS

Among the 186 patients who underwent PHCF for symptomatic PLD, 170 (91.4%) patients had ADPKD and 16 (8.6%) had autosomal dominant polycystic liver disease. The majority of patients were women (90.5%) and Caucasian (91.8%). Mean age at operation was 49 ± 9.6 years. Mean number of hepatic segments resected per patient was 4.42 ± 1.2. At the time of PHCF, 38%