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Liver Transplantation for Primary Sclerosing Cholangitis: A 20-Year Single-Center Experience

Primer Sklerozan Kolanjit İçin Karaciğer Nakli: 20 Yıllık Tek Merkez Deneyimi

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Liver Transplantation for Primary Sclerosing Cholangitis: A 20-Year Single-Center Experience

ABSTRACT

Objective: Primary sclerosing cholangitis is a chronic, progressive cholestatic liver disease that frequently leads to liver transplantation. Although liver transplantation remains the only definitive treatment for advanced disease, post-transplant recurrence may adversely influence long-term outcomes. This study aimed to evaluate recurrence rates and clinical outcomes after liver transplantation in patients with primary sclerosing cholangitis.

Material and Methods: This retrospective single-center study included 37 patients who underwent liver transplantation between December 2004 and April 2025 due to primary sclerosing cholangitis. Demographic characteristics, clinical findings, surgical approaches, medical treatments, complications, recurrence patterns, and survival outcomes were evaluated using descriptive statistical methods.

Results: The cohort consisted of sixteen males and twenty-one females with a mean age of 41.9 years at the time of transplantation. The mean disease duration was 6.1 years, and the mean Model for End-Stage Liver Disease score was 18.4. Inflammatory bowel disease was present in 43 % of patients, all diagnosed with ulcerative colitis. Living donor liver transplantation was performed in thirty-six patients, while one patient received a deceased donor graft. Incidental cholangiocarcinoma was identified in two explanted livers. During follow-up, disease recurrence developed in nine patients at a mean of 45.6 months, and two patients required re-transplantation. Five-year mortality was 13.5 percent, with three deaths associated with recurrence and related complications.

Conclusion: Liver transplantation provides favorable long-term survival in patients with primary sclerosing cholangitis; however, recurrence remains a significant challenge. Structured long-term surveillance, optimized medical management, and multidisciplinary follow-up are essential to improve post-transplant outcomes.

Keywords: Cholangitis, Cholestasis, Liver transplantation.

Primer Sklerozan Kolanjit İçin Karaciğer Nakli: 20 Yıllık Tek Merkez Deneyimi

ÖZ

Amaç: Primer sklerozan kolanjit, sıklıkla karaciğer nakline yol açan kronik ve progresif kolestatik bir karaciğer hastalığıdır. Karaciğer nakli ileri evre hastalık için tek kesin tedavi seçeneği olmakla birlikte, nakil sonrası hastalık nüksü uzun dönem sonuçları olumsuz etkileyebilmektedir. Bu çalışmanın amacı, primer sklerozan kolanjit nedeniyle karaciğer nakli uygulanan hastalarda nüks oranlarını ve nakil sonrası klinik sonuçları değerlendirmektir.

Gereç ve Yöntem: Bu retrospektif tek merkezli çalışmaya, Aralık 2004 ile Nisan 2025 tarihleri arasında primer sklerozan kolanjit nedeniyle karaciğer nakli yapılan 37 hasta dâhil edilmiştir. Demografik özellikler, klinik bulgular, cerrahi yaklaşımlar, medikal tedaviler, komplikasyonlar, nüks paternleri ve sağkalım sonuçları tanımlayıcı istatistiksel yöntemler kullanılarak değerlendirilmiştir.

Bulgular: Çalışma kohortu, nakil sırasında ortalama yaşı 41,9 yıl olan on altı erkek ve yirmi bir kadından oluşmaktaydı. Ortalama hastalık süresi 6,1 yıl, ortalama Model for End-Stage Liver Disease skoru ise 18,4 olarak saptandı. Hastaların %43'ünde inflamatuvar bağırsak hastalığı mevcuttu ve tümü ülseratif kolit tanısı almıştı. Otuz altı hastaya canlı vericili, bir hastaya ise kadavra vericili karaciğer nakli uygulandı. Eksplante edilen karaciğerlerin ikisinde insidental kolanjiyokarsinom saptandı. İzlem süresince dokuz hastada ortalama 45,6 ayda hastalık nüksü gelişti ve iki hasta yeniden nakil gerektirdi. Beş yıllık mortalite yüzde 13,5 olup ölümlerin üçü hastalık nüksü ve ilişkili komplikasyonlara bağlıydı.

Sonuç: Primer sklerozan kolanjitli hastalarda karaciğer nakli uzun dönem sağkalım açısından etkili bir tedavi seçeneği sunmakla birlikte hastalık nüksü önemli bir sorun olmaya devam etmektedir. Yapılandırılmış uzun dönem izlem, optimize edilmiş medikal tedavi ve multidisipliner yaklaşım nakil sonrası sonuçların iyileştirilmesi için gereklidir.

Anahtar Sözcükler: Karaciğer nakli, Kolanjitis, Kolestaz.

Introduction

Primary sclerosing cholangitis (PSC) is a chronic, progressive inflammatory disease characterized by destruction and inflammation of the intrahepatic and/or extrahepatic bile ducts. Although its etiology remains unclear, it is considered to be multifactorial. The disease is associated with an increased risk of fibrosis, liver failure, cirrhosis, and malignancy (1-3). Primary sclerosing cholangitis is a rare chronic cholestatic liver disease, with a reported worldwide prevalence of approximately 0.5–16.2 per 100,000 individuals and an annual incidence of 0.2–1.3 per 100,000 individuals (4).

The most widely accepted pathogenic hypothesis suggests that immune-mediated injury to cholangiocytes, potentially triggered by toxins or infectious agents, results in progressive bile duct destruction, chronic cholestasis, and fibrosis (5). Clinically, PSC may be asymptomatic or present with fatigue, weight loss, pruritus, jaundice, and right upper quadrant abdominal pain (6). Histologically, periductal fibrosis, bile duct proliferation, and ductopenia are observed in association with inflammation.

Although PSC exhibits features typical of autoimmune diseases, its response to immunosuppressive therapies is limited. Although ursodeoxycholic acid (UDCA) may be administered in selected patients, no definitive curative medical therapy exists. Therefore, liver transplantation remains the only effective treatment option for advanced-stage disease, significantly improving both survival and quality of life. However, in a subset of PSC patients, the disease may recur after transplantation. Post-transplant recurrence of PSC may significantly affect patient prognosis and subsequent treatment strategies. As part of the autoimmune disease spectrum, PSC is commonly associated with inflammatory bowel disease (IBD) (7).

This study evaluates the management strategies, complications, and post-transplant outcomes of patients undergoing liver transplantation for PSC.

Material and Method

Approval for the study was obtained from the İstanbul Demiroğlu Bilim University Faculty of Medicine Research Ethics Committee on 29/01/2025. Decision no: 51016662/44696. The study was conducted in accordance with the Declaration of Helsinki. The requirement for informed consent was waived due to the retrospective design of the study.

This study is a retrospective, single-center analysis conducted to evaluate the outcomes of patients who underwent liver transplantation for primary sclerosing cholangitis (PSC) among a total of 1,652 liver transplantations performed between December 2004 and April 2025. Patients who underwent liver transplantation at our center during this 20-year period and had a confirmed diagnosis of PSC were included in the study.

Patients with an unconfirmed diagnosis of PSC prior to transplantation, those with incomplete clinical, laboratory, or follow-up data, and those whose early post-transplant follow-up data (within the first 30 days) were unavailable were excluded from the analysis.

Overall, 37 patients (2.2%) were identified as having PSC as the underlying etiology for liver transplantation.

Data Collection

Demographic characteristics (age and sex), age at PSC diagnosis, age at transplantation, disease duration, Model for End-Stage Liver Disease (MELD) scores, pre-transplant complications, and comorbidities were retrospectively obtained from patient medical records and electronic hospital databases.

During the pre-transplant period, clinical conditions including recurrent cholangitis episodes, portal hypertension–related complications (esophageal variceal bleeding and ascites), hepatic encephalopathy, and hepatorenal syndrome were recorded. In addition, imaging findings were reviewed for the presence of hepatocellular carcinoma (HCC) and cholangiocarcinoma (CCA). A history of IBD was also documented.

Pre-transplant Evaluation

All patients underwent a comprehensive pre-transplant evaluation before listing for liver transplantation. The assessment included detailed clinical examination, laboratory investigations, and radiological imaging to evaluate liver disease severity and potential extrahepatic complications. Upper gastrointestinal endoscopy was performed to assess the presence of portal hypertension and varices. Colonoscopy was routinely carried out to screen for inflammatory bowel disease and colorectal neoplasia, which are known to be associated with primary sclerosing cholangitis. Additional imaging studies were used when necessary to exclude hepatobiliary malignancies and to evaluate biliary tract pathology.

Transplantation Procedure

Liver transplantation was performed using either living donor grafts (right or left lobe) or deceased donor grafts. Recipient hepatectomy was carried out through a reverse-T incision using the piggy-back technique with preservation of the inferior vena cava. Following removal of the native liver, the donor graft was implanted. Vascular reconstruction was performed with end-to-end anastomoses of the hepatic vein(s), portal vein, and hepatic artery using standard techniques.

Biliary reconstruction was achieved either by duct-to-duct anastomosis or by Roux-en-Y hepaticojejunostomy depending on recipient and donor characteristics. The type of graft used at transplantation (living donor right lobe, left lobe, or deceased donor graft), the biliary reconstruction technique, and the degree of donor–recipient relationship were recorded. Explant liver pathology reports were reviewed to assess the presence of malignancy.

Post-Transplant Follow-up

Patients were followed from the date of liver transplantation until death, re-transplantation, or the last clinical follow-up. Follow-up data were obtained from outpatient clinic visits and hospital records. Post-transplant immunosuppressive regimens and modifications made during follow-up were analyzed. The occurrence of acute rejection, the need for immunosuppression modification, recurrence of PSC, requirement for re-transplantation, and time to disease recurrence were recorded. In addition, the development of de novo inflammatory bowel disease in the post-transplant period was evaluated.

Recurrence of PSC was diagnosed based on clinical findings, cholestatic biochemical abnormalities, and characteristic biliary imaging findings on magnetic resonance cholangiopancreatography (MRCP), after exclusion of other identifiable causes of biliary strictures such as anastomotic stricture, hepatic artery thrombosis, or secondary sclerosing cholangitis.

Statistical Analysis

Statistical analysis was performed using SPSS version 27 (IBM Corp., Armonk, NY, USA). Descriptive statistical methods were used in this study. Continuous variables were expressed as mean ± standard deviation (minimum–maximum), and categorical variables were presented as numbers and percentages. No comparative analyses or advanced statistical tests were performed. Since the study was purely descriptive in design and did not involve hypothesis testing or group comparisons, p-values were not calculated or reported.

Results

A total of 37 patients were included in the study, of whom 16 (43.2%) were male (Table I). The mean age at the time of PSC diagnosis was 36.57 ± 15.47 years (range, 5–63), and the mean age at liver transplantation was 41.92 ± 14.14 years (range, 16–69). The mean disease duration between diagnosis and transplantation was 6.15 ± 4.42 years (range, 1–17). The mean Model for End-Stage Liver Disease (MELD) score at the time of transplantation was 18.38 ± 6.40 (range, 8–32). The median follow-up duration was 5 years (range, 1-12 years).

Table I. Baseline Demographic and Clinical Characteristics

Variable	Value
Number of patients	37
Male sex, n (%)	16 (43.2)
Age at PSC diagnosis, years	36.57 ± 15.47 (5–63)
Age at transplantation, years	41.92 ± 14.14 (16–69)
Disease duration before LT, years	6.15 ± 4.42 (1–17)
MELD score at LT	18.38 ± 6.40 (8–32)
Ulcerative colitis, n (%)	16 (43.2)

When pre-transplant complications were evaluated, recurrent cholangitis episodes were observed in 17 patients, esophageal variceal bleeding in 4 patients, and ascites in 12 patients. In addition, other complications, including hepatic encephalopathy and hepatorenal syndrome were present in 12 patients. No hepatocellular carcinoma or cholangiocarcinoma was detected on preoperative imaging in any patient. All patients were receiving ursodeoxycholic acid (UDCA) therapy in the pre-transplant period (Table II).

Table II. Pre-transplant Complications

Complication	n (%)
Recurrent cholangitis	17 (45.9)
Esophageal variceal bleeding	4 (10.8)
Ascites	12 (32.4)
Hepatic encephalopathy	12 (32.4)
Pre-transplant HCC	0 (0)
Pre-transplant cholangiocarcinoma	0 (0)

Azathioprine was used in two patients, and a combination of steroids and azathioprine was administered in two patients. Sixteen patients (43.2%) had a history of inflammatory bowel disease, and all of these patients were diagnosed with ulcerative colitis.

During the perioperative period, a left lobe graft was used in 1 patient, a right lobe graft in 35 patients, and a deceased donor graft in 1 patient. Biliary reconstruction was performed as hepaticojejunostomy in 18 patients and as duct-to-duct anastomosis in 19 patients. Among patients who underwent living donor liver transplantation, the donor was a first-degree relative in 14 cases. Incidental cholangiocarcinoma was identified in 2 patients on explant pathology (Table III).

Table III. Transplant Characteristics and Post-transplant Outcomes

Variable	n (%)
Transplant and Operative Characteristics	
Right lobe graft	35 (94.6)
Left lobe graft	1 (2.7)
Deceased donor graft	1 (2.7)
Hepaticojejunostomy	18 (48.6)
Duct-to-duct anastomosis	19 (51.4)
First-degree related donor	14 (37.8)
Incidental cholangiocarcinoma in explant	2 (5.4)
Post-transplant Outcomes	
Immunosuppression modification	4 (10.8)
Acute rejection	6 (16.2)
PSC recurrence	9 (24.3)
Re-transplantation	2 (5.4)
De novo inflammatory bowel disease	1 (2.7)
5-year mortality	5 (13.5)

In the post-transplant period, the most commonly used immunosuppressive regimen consisted of steroids, tacrolimus, and mycophenolate mofetil; cyclosporine was used instead of tacrolimus in two patients. During follow-up, 4 patients (10.8%) required modification of the immunosuppressive regimen. All patients continued ursodeoxycholic acid (UDCA) therapy in the post-transplant period. Acute rejection occurred in 6 patients (16.2%) and was successfully managed with immunosuppression modification; no graft loss due to rejection was observed.

During follow-up, recurrence of PSC was detected in 9 patients (24.3%), and 2 of these patients required re-transplantation. The mean time from transplantation to recurrence was 45.57 ± 31.79 months (range, 6–96). De novo inflammatory bowel disease presenting as pancolitis developed in one patient 60 months after transplantation.

During the 5-year follow-up period, a total of 5 patients (13.5%) died. Two deaths were due to extrahepatic causes, whereas three deaths

were attributed to disease recurrence and related complications.

Discussion

In the United States, end-stage liver disease due to cholestatic liver diseases accounts for approximately 8% of adult liver transplantations (8). Among these conditions, primary sclerosing cholangitis (PSC) represents an important indication for liver transplantation. In patients with PSC, current indications for liver transplantation include not only MELD score–based indications such as decompensated cirrhosis, but also PSC-specific indications eligible for MELD exception points, including early-stage hilar cholangiocarcinoma, recurrent bacterial cholangitis, and intractable pruritus. In our cohort, 17 patients (45.9%) underwent liver transplantation due to refractory cholangitis. Liver transplantation significantly improves both survival and quality of life in patients with PSC. However, biliary complications, disease recurrence, and the risk of malignancy remain major post-transplant concerns.

Recipients of living donor liver transplantation (LDLT) have been reported to have higher rates of disease recurrence and biliary complications compared with recipients of deceased donor liver transplantation (8). In our series, only one patient underwent deceased donor liver transplantation; therefore, a comparative analysis between living donor and deceased donor transplantation was not feasible. Close and regular post-transplant follow-up plays a critical role in improving long-term outcomes in this patient population.

PSC is a disease of middle age (typically 35–51 years) and is associated with an increased risk of hepatobiliary malignancies, particularly CCA (6, 9, 10). CCA occurs in approximately 2–6% of cases (11). In our study, incidental CCA without clinical or radiological manifestations was identified in 2 patients (5.4%). In our cohort, which also included patients diagnosed during the pediatric period, the mean age at diagnosis was consistent with that reported in the literature.

Although PSC is known to be more common in males, with a reported male-to-female ratio of approximately 7:1, female sex predominated in our cohort. In PSC, inflammatory bowel disease accompanies approximately 50–85% of cases, with ulcerative colitis reported in 70–80% and Crohn's disease in 7–21% of patients; therefore, an increased risk of colorectal cancer in this population should be considered (7, 12-14). In this study, inflammatory bowel disease was present in 43.2% of patients, and all cases were ulcerative colitis; no patients had Crohn's disease or indeterminate colitis. Previous studies have reported that de novo inflammatory bowel disease may develop in approximately 14–30% of PSC patients within 10 years after liver transplantation (5, 15). In our cohort, de novo ulcerative colitis developed in only one patient, which was lower than the rates reported in the literature.

Although the efficacy of ursodeoxycholic acid (UDCA) in the treatment of PSC has not been clearly established, some studies recommend its use, whereas others do not support routine administration (7, 16). In our study, UDCA therapy was used in all patients during both the pre- and post-transplant periods.

Although recurrence rates of PSC after liver transplantation are low during the first year, they have been reported to range between 20% and 30% during long-term follow-up (7). In our study, PSC recurrence was observed in 24.3% of patients, and graft loss occurred in two patients due to disease recurrence. Careful and close follow-up is essential to facilitate early detection (17, 18).

The diagnosis of recurrent PSC is challenging, as non-anastomotic biliary strictures may nonspecifically affect many liver grafts. Therefore, other potential causes of biliary strictures must be excluded before establishing a diagnosis of recurrence. Donor and recipient demographic characteristics, use of extended-criteria donors, presence of active inflammatory bowel disease, and episodes of acute rejection have been identified as predisposing risk factors for recurrence (19).

Recurrent PSC may be diagnosed based on progressive biliary strictures on cholangiography and/or histological findings consistent with PSC occurring more than 90 days after liver transplantation, after exclusion of other identifiable causes (7). There is currently no effective prophylactic strategy to prevent recurrence of PSC (8). The median time from transplantation to recurrent PSC has been reported to be approximately 5 years (7). In our study, recurrence occurred at a mean of 45 months (approximately 4 years). In light of these findings, a multidisciplinary approach and careful management of immunosuppressive therapy are recommended to reduce the risk of recurrence. Regular follow-up during this period is essential to facilitate early diagnosis and implementation of effective treatment strategies, thereby mitigating the impact of disease recurrence.

In liver transplantation for PSC, duct-to-duct biliary anastomosis was historically avoided due to concerns regarding the potential development of cholangiocarcinoma in the residual bile duct and an increased risk of biliary strictures related to the underlying PSC. However, previous studies have demonstrated that, compared with hepaticojejunostomy, duct-to-duct anastomosis is associated with comparable patient and graft survival, similar rates of PSC recurrence, minimal risk of cholangiocarcinoma, and comparable risks of biliary strictures and cholangitis. The choice of biliary reconstruction technique is therefore largely at the discretion of the transplant surgical team (8).

Nevertheless, duct-to-duct anastomosis is generally recommended as the preferred approach when Roux-en-Y hepaticojejunostomy is not required (7). In this study, the distribution of biliary reconstruction techniques was nearly equal between duct-to-duct anastomosis and hepaticojejunostomy.

The main limitations of this study include its retrospective design and the relatively small sample size. Nevertheless, our findings provide important insights into the natural course of PSC in the post-transplant setting. Optimization of immunosuppressive regimens and close clinical follow-up are of critical importance to reduce the risk of disease recurrence.

Conclusion

Early diagnosis in PSC, appropriate management of complications, and optimal timing of liver transplantation are crucial components of patient care. Liver transplantation due to PSC offers an effective therapeutic option for patients with advanced-stage disease. However, in these patients, the risk of disease recurrence and the management of other post-transplant complications require meticulous planning. Long-term follow-up and a multidisciplinary approach can significantly enhance treatment outcomes.

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