# Partial hepatectomy is curative for the localized type of Caroli's disease: A case report and review of the literature

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**The case of a 58-year-old woman who was diagnosed with the localized type of Caroli's disease is presented. This disease involves the whole of the left half of the liver. A left hepatectomy was followed by complete resolution of symptoms. The current article suggests that hepatic resection may be aggressively performed in selected patients with the localized form of Caroli's disease**

***Keywords: Caroli's disease, surgical treatment, hepatectomy***
***Surgeon, 1 April 101-105***

## INTRODUCTION

Caroli's disease is defined as a congenital disease characterized by cystic dilatations of the intrahepatic biliary tree.[1,2] The prognosis of Caroli's disease is dependent on the extent of the lesions. If limited to one segment, one sector, or one half of the liver it may be excised radically and effectively by corresponding removal of the hepatic parenchyma.[3-11] If involving the whole of the duct system of the hepatic parenchyma, it is associated with a grave prognosis, regardless of the therapeutic methods used.[12,13]

We report a patient with the localized type of Caroli's disease which involved the whole of the left half of the liver and in whom left hepatectomy was followed by complete resolution of the symptoms. We also review the previously published cases of localized Caroli's disease treated by partial hepatectomy.

## CASE REPORT

A 58-year-old woman was admitted on June 27, 2003, with a seven-day history of fever, chills and right upper quadrant abdominal pain. On physical examination, the right subcostal region was tender. She had a white blood cell count of 10200/mm3, aspartate aminotransferase of 53 U/L, lanine aminotransferase of 142 U/L, alkaline phosphatase of 605 U/L, gamma glutamyl transpeptidase of 408 U/L and a total/direct bilirubin of 2.2/1.5 mg/dL.

Abdominal ultrasound showed stones in the cystic dilatations of the left intrahepatic biliary tract and in the gallbladder. The common bile duct was 13mm in diameter. Computed tomography (CT) of the abdomen after the first endoscopic retrograde cholangiopancreatography (ERCP) confirmed that cystic cavities containing air were present in the left half of the liver (Figure 1). These findings were suggestive of the localized type of Caroli's disease. Three subsequent ERCPs done at seven-day intervals confirmed the dilatations in the left intrahepatic biliary system (Figure 2). In the first ERCP, a sphincterotomy was performed and a mildly dilated common bile duct was seen. However the intrahepatic biliary tract could be not reached. In the second attempt, the stones in the common bile duct were seen and removed with the basket. Additional ERCP findings included stones also in the left intrahepatic biliary tract but it was not possible to extract these stones with the balloon and the basket. A nasobiliary drain was placed in the common bile duct because of cholangitis. In the last attempt, extraction of the stones failed after removing the nasobiliary

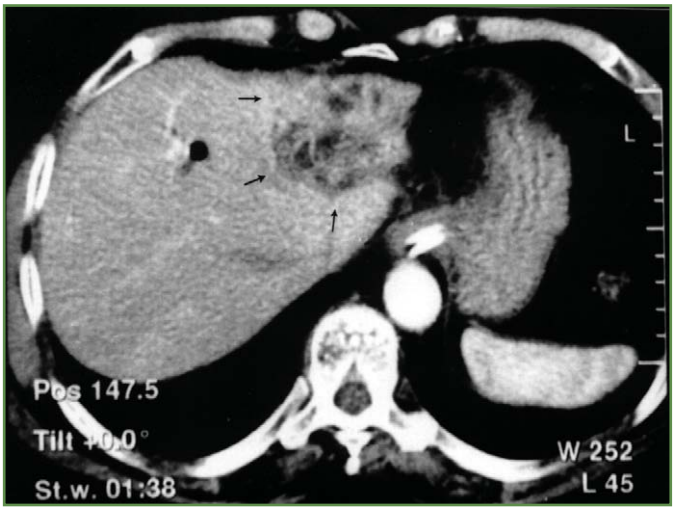


***Figure 1:*** *The CT scan of the abdomen showing large cystic cavities in the left half of the liver marked with arrows.*

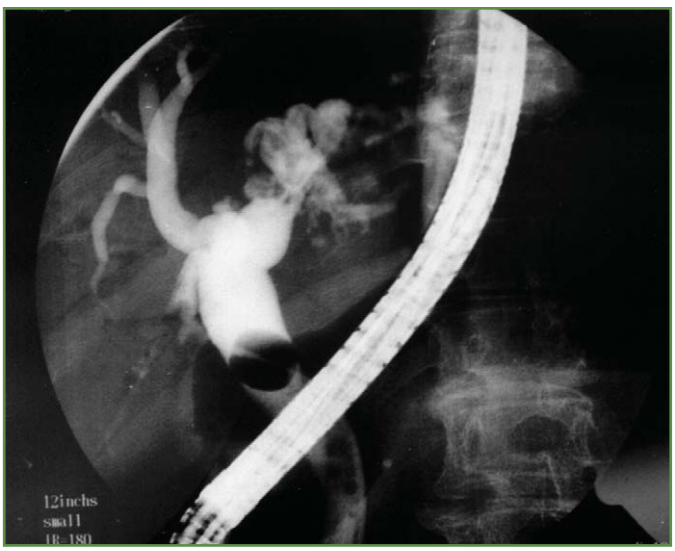


***Figure 2:*** *ERCP showing cystic dilatation of the left intrahepatic biliary system.*

drain. Therefore, the nasobiliary drain was inserted again and extracorporeal shockwave lithotripsy (ESWL) was planned but not carried out. The patient underwent an elective laparotomy on July 25, 2003 with an initial diagnosis of the localized type of Caroli's disease. At laparotomy, cystic dilatations of the bile ducts were seen and palpated in segments two, three and four of the liver. The remainder of the liver had a normal appearance. A left hepatectomy including the segment one and cholecystectomy were performed. In addition, the stones in the common bile duct were removed via choledochotomy and a T-tube was placed in the common bile duct.

Macroscopic examination of the resected left liver revealed numerous cysts of various sizes communicating with the intrahepatic bile ducts. Many of these cysts contained one to several dark stones (Figure 3). Microscopic examination of the resected hepatic tissue showed markedly dilated segmental bile ducts, parenchymal and periductal fibrosis, cholangitis in the portal areas, porto-portal fibrotic bridging, moderate dysplasia of the biliary epithelium and portal triads infiltrated by polymorphonuclear cells.

Post-operative recovery was uneventful apart from a wound infection which settled rapidly with open wound care. A T-tube cholangiogram post-operatively showed mild leakage of bile from the cut surface of the resected liver which ceased spontaneously (Figure 4). The T-tube was removed after a month. From the day of surgery to May 2004, the patient's condition remained excellent. Follow-up ultrasound and CT of the liver performed in May 2004 revealed a normal remaining right liver (Figure 5). On follow-up ERCP, right and extrahepatic bile ducts were normal (Figure 6). When last seen in October 2004, she was well.

## DISCUSSION

There are two types of Caroli's disease. In the diffuse or total type, cystic dilatation of the segmental bile ducts affects the whole intrahepatic biliary tree of the liver. In the localized or limited type, cystic dilatation of the segmental bile ducts is confined to one segment, one sector, or one half of the liver. The question of which type of Caroli's disease is more frequently seen is controversial. The localized form which at most involves the entire left lobe of the liver, is the most frequent form according to Mercaider *et al* (1984).[3] But Ramond *et al* (1984)

**TABLE 1. REPORTED CASES OF THE LOCALIZED TYPE OF CAROLI'S DISEASE**

| Localization | | Number of Cases | Extent of hepatic resection | Reference |
|---|---|---|---|---|
| Right liver | | | | |
| | Whole | 2 | Right hepatectomy | 4, 21 |
| | Segment 7 | 1 | Right hepatectomy | 4 |
| | Segment 6 and 7 | 1 | Segment 6 and 7 resection | 7 |
| Right liver and segment 4 | | 2 | Right lobectomy | 5, 8 |
| Left liver | | | | |
| | Whole | 14 | Left hepatectomy* | 1, 2, 3, 4, 6, 9, 10, 15, 17, 19, 21 |
| | Segment 2 and 3 | 10 | Left lobectomy | 3, 4, 11, 14, 16, 18, 20 |
| Left liver and segment 6 and 7 | | 1 | left hepatectomy | 22 |

* Left lobectomy and Hepp's operation in one case (3)

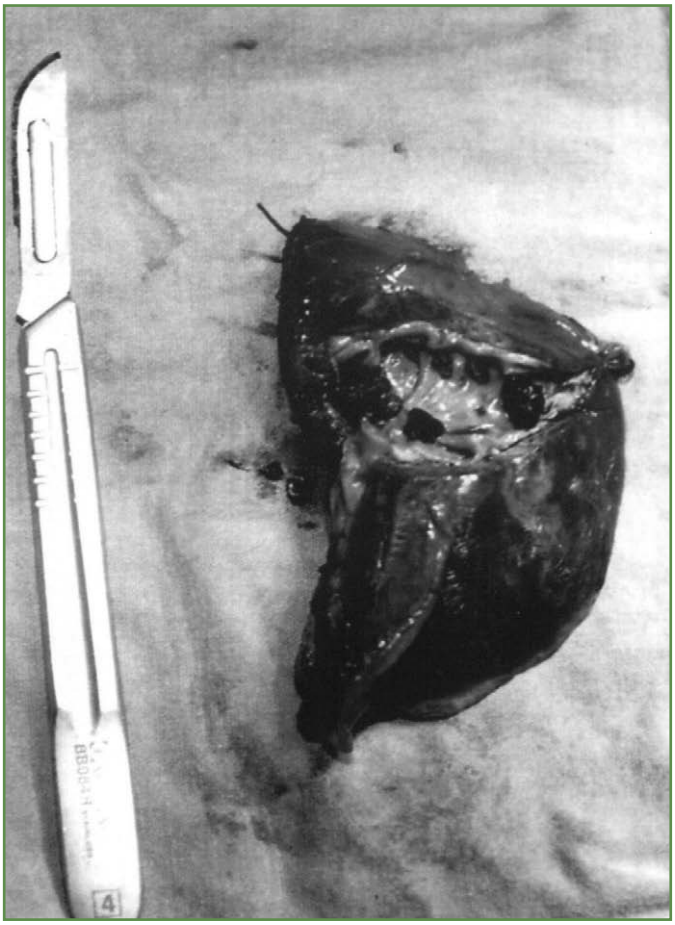

***Figure 3:*** *Resected specimen, left hepatectomy. There were cystic dilatations of the bile ducts in the specimen. The cysts were filled with stones.*

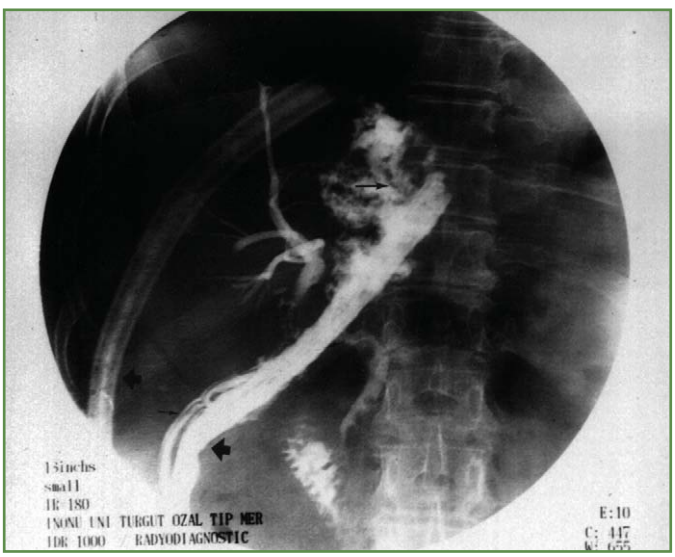


***Figure 4:*** *T-tube (short arrow) cholangiogram at ten days post-operatively demonstrating mild bile leakage (long arrow) from the cut surface of the liver. Note absence of the left ductal system. Thick arrows show Jackson-Pratt drains.*

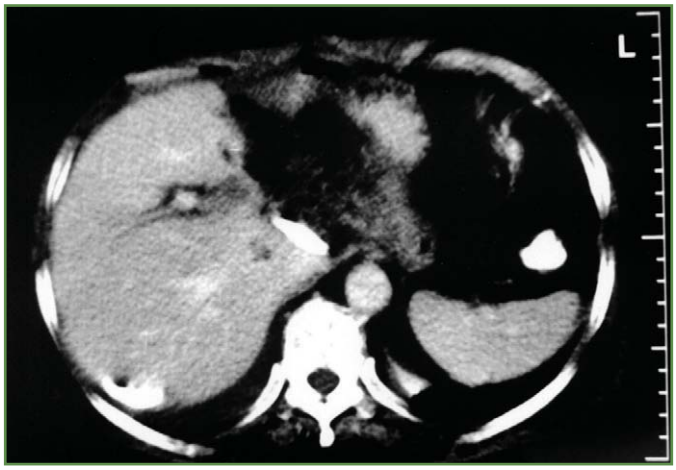

***Figure 5:*** *The CT scan of the abdomen ten months after partial hepatectomy showing the remnant normal liver.*

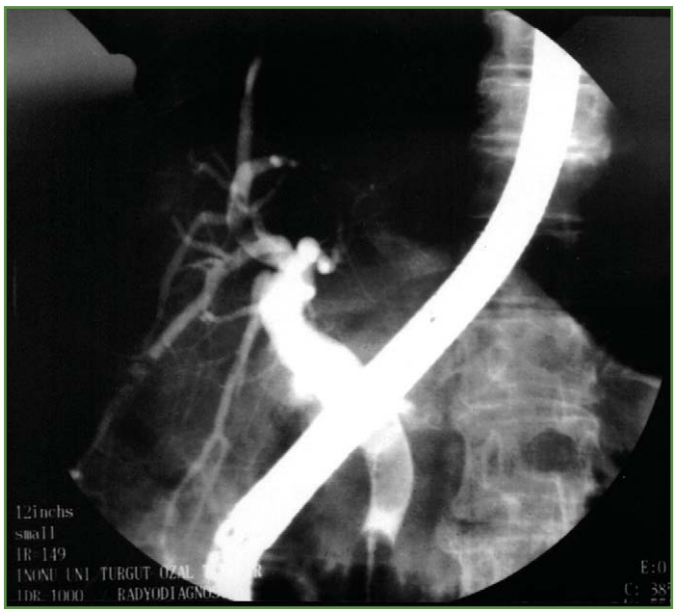


***Figure 6:*** *ERCP ten months after partial hepatectomy demonstrating normal right ductal system.*

found that the diffuse type was more common than the localized type.[5] From the therapeutic standpoint, a distinction must be made between limited and total forms of Caroli's disease. In the present report, a rare clinical case of segmental dilatation of the intrahepatic biliary tree confined to the left hemi-liver, is described.

The localized forms, which involve the whole of the left half of the liver, or the right-half of the liver, are curable by surgery. They should be treated by hemi-hepatectomy with associated treatment of any problem affecting the common duct.[3,6] As in our case, most patients also need common duct surgery. The incidence of cases suitable for hepatic resection is fairly low. The features of the previously reported thirty one cases of the

localized type of Caroli's disease treated by partial hepatectomy are presented in Table 1. Of the 32 cases with localized Caroli's disease, including our case, 25 were localized to the left half of the liver and six to the right half of the liver.[1-11,14-21] However, the lesions extended to segment four from the right hemi-liver in the cases reported by Ramond *et al* (1984) and Kumar *et al* (2001).[5,8] Of 25 cases localized to the left half of the liver, ten were solely in segments.[2,3] One case had localized lesions in both sides of the liver.[22] Compared with right hepatectomy, a longer disease-free period has been obtained after a resection of segment two, three (left hepatic lobectomy) for the treatment of the localized type of Caroli's disease (45.4 months for left hepatic lobectomy, 23.6 months for left hepatectomy and 20.2 months for right hepatectomy). However, these figures are related to the time of follow-up in the cases reported. It is also possible that an improved chance of cure could be achieved with accurate disease localization and complete resection, than if diseased liver were left behind as reported by Guivarc'h *et al* (1982).[16]

The localized type of Caroli's disease and congenital hepatic fibrosis are not frequently associated as they are in this case. However the diffuse type is usually associated with congenital hepatic fibrosis, characterised by bile duct proliferation and malformation. The main consequence of congenital hepatic fibrosis is portal hypertension and oesophageal varices. In the light of the natural history of Caroli's disease (cirrhosis, variceal bleeding and liver failure) hepatic transplantation offers the best chance of success. Orthotopic liver transplantation has been successfully employed for Caroli's disease.[23]

Biliodigestive anastomoses or endoscopic sphincterotomy are often unsuccessful in treating and preventing complications such as intraductal lithiasis, repeated cholangitis and liver abscesses in the localized form of Caroli's disease.[4,7,24] Considering these problems, resection of the diseased segment or lobe of the liver is apparently advantageous. The incidence of cases suitable for hepatic resection may be low, but the most accurate and reliable treatment is undoubtedly removal of the diseased portion of the liver if possible, as suggested by several investigators.[3-5,7-11] Left hepatectomy was chosen in our patient because the cholangitis and the intrahepatic stones were not controlled by repeated ERCPs and endoscopic sphincterotomy. Surgery was followed by complete cessation of cholangitis. Attention should be given to the complete resection of the liver parenchyma including the stones, which can be extensive in these patients.[3] Progression of the disease to the remaining liver may be possible and this situation should be managed by liver transplantation.[25] Although a more extensive resection with hepaticojejunostomy is occasionally possible in patients who present with diffuse form of Caroli's disease, cholangitis attacks due to the disease in the remaining liver and the considerable risk of malignant transformation are major considerations.[26] However, cholangiocarcinoma has also been reported in a patient with localized form of Caroli's disease.[24]

## CONCLUSION

This case indicates that hepatic resection may be aggressively performed in selected patients with the localized form of Caroli's disease. This procedure will most probably remove both the stones and the potential site of malignant change.



## REFERENCES


1. Caroli J, Coinaud C, Soupault R, Porcher P, Eteve J. Une affection nouvelle, sans doute congénitale, des voies biliaires: La dilatation kystique unilobaire des canaux hépatiques. *Sem Hop Paris* 1958; **34**: 136-42.
2. Caroli J, Laverdant C, Moine D, Hepp J, Hadchhouel P. La dilatation kystique unilobaire des canaux biliaires segmentaires (maladie de J. Caroli) avec empierrement microcalculeux. *Med Chir Dig* 1974; **3**: 371-75.
3. Mercadier M, Chigot JP, Clot JP, Langlois P, Lansiaux P. Caroli's disease. *World J Surg* 1984; **8**: 22-29.
4. Nagasue N. Successful treatment of Caroli's disease by hepatic resection. Report of six patients. *Ann Surg* 1984; **200**: 718-23.
5. Ramond MJ, Huguet C, Danan G, Rueff B, Benhamou JP. Partial hepatectomy in the treatment of Caroli's disease. Report of a case and review of the literature. *Dig Dis Sci* 1984; **29**: 367-70.
6. Roda E, Sama C, Festi D, Aldini R, Mazzella G, Roda A, Barbara L. Caroli's disease. *Am J Gastroenterol* 1979; **71**: 621-26.
7. Sandle GI, Lodge JPA. A 32-year-old man with recurrent cholangitis. *Lancet* 1997; **350**: 408.
8. Kumar KS, Nair S, Hertan HI, Grover H. Isolated intrahepatic biliary dilatation in a patient with acquired immune deficiency syndrome (AIDS): AIDS cholangiopathy versus incidental unilobar Caroli's disease. *J Clin Gastroenterol* 2001; **32**: 79-81.
9. Tamiolakis D, Arvanitidou V, Nikolaidou S, Barbagadaki S, Avgidou K, Boglou P, Papadopoulos N. Caroli's syndrome. A case report and review of the literature. *Minerva Gastroenterol Dietol* 2004; **50**: 179-81.
10. Joshi RM, Wagle PK, Darbari A, Chhabra DG, Patnaik PS, Katrak MP. Hepatic resection for benign liver pathology-report of two cases. *Indian J Gastroenterol* 2002; **21**: 157-59.
11. Giovanardi RO. Monolobar Caroli's disease in an adult. Case report. *Hepatogastroenterology* 2003; **50**: 2185-87.
12. Dayton MT, Longmire WP, Tompkins RK. Caroli's disease: A premalignant condition. *Am J Surg* 1983; **145**: 41-48.
13. Witlin LT, Gadacz TR, Zuidema GD, Kridelbaugh WW. Transhepatic decompression of the biliary tree in Caroli's disease. *Surgery* 1982; **97**: 205-9.
14. Borda F, Uribarrena R, Puras A, Ortiz H, Rivero-Puente A. Résection hépatique pour maladie de Caroli limitée au lobe gauche. *Gastroenterol Clin Biol* 1982; **6**: 511-12.
15. Gillet, cited by Colin R, Vayre P. Lithiase intrahépatique et ses migrations. Paris, Masson 1979.
16. Guivarc'h M, Roullet-Audy JC, Amstrong O, Barbagelatta M. Maladie de Caroli localisée au lobe gauche. A propos d'une nouvelle observation. *Med Chir Dig* 1982; **11**: 263-67.
17. Leroy JP, Griffe J, Briere J, Volant A, Le Menn G. Un nouveau cas de carcinome développé sur maladie de Caroli. *Sem Hop Paris* 1982; **58**: 417-19.
18. Mouiel J, Bourgeon R. La lithiase biliaire intrahépatique. A propos de 39 observations personnelles. *Chirurgie* 1979; **105**: 437-49.

19. Roudot-Thoraval F, Zafrani ES, Hannoun S, Fagniez PL, Metreau JM, Dhumeaux D. Résection hépatique pour maladie de Caroli limitée au foie gauche. Découverte fortuite d'un cholangiocarcinome. *Gastroenterol Clin Biol* 1983; **7**: 319-20.
20. Schrumpf E, Bergan A, Blomhoff JP, Hoel PS, Kolmannskog F. Partial hepatectomy in Caroli's disease. *Scand J Gastroenterol* 1981; **16**: 581-86.
21. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cyst. Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. *Am J Surg* 1977; **134**: 263-69.
22. Farah M, Helou E, Nasser W, Tabbara W. Un nouveau cas de maladie de Caroli avec dilatations géantes bilobaires segmentaires, compliquée de lithiase et guérie chirurgicalement. *Rev Med Chir Mal Foie* 1966; **41**: 305-28.
23. Ulrich F, Steinmuller T, Settmacher U, Muller AR, Jonas S, Tullius SG, Neuhaus P. Therapy of Caroli's disease by orthotopic liver transplantation. *Transplant Proc* 2002; **34**: 2279-80.
24. Abdalla EK, Forsmark CE, Lauwers GY, Vauthey JN. Monolobar Caroli's disease and cholangiocarcinoma. *HPB Surg* 1999; **11**: 271-77.
25. Ammori BJ, Jenkins BL, Lim PCM, Prasad KR, Pollard SG, Lodge PA. Surgical strategy for cystic diseases of the liver in a Western hepatobiliary center. *World J Surg* 2002; **26**: 462-69.
26. Aeberhard P. Surgical management of Caroli's disease involving both lobes of the liver. *Br J Surg* 1985; **72**: 651-52.

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