

# Polycystic Liver Disease Treated by Fenestration and Segmental Liver Resection

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## Summary

A case of symptomatic polycystic liver disease treated by fenestration and segmental liver resection is reported. The intraoperative use of ultrasound to define the plane of liver resection is emphasised. No significant post-operative complication was encountered. The clinical presentation, management and treatment are discussed.

**Key words:** Polycystic liver disease, Fenestration, Segmental liver resection.

## Introduction

Polycystic liver disease (PLD) is a rare clinical entity which is now seen more frequently due to the wider application of non-invasive radiological imaging procedures such as ultrasound and CT scan. The prevalence of PLD associated with autosomal dominant polycystic kidney (ADPKD) has been well documented at approximately 50 per cent. Advances in long term maintenance haemodialysis and improved medical treatment have prolonged life in patients with severe degrees of renal impairment from ADPKD. Subsequently, the number of cases associated with PLD has also increased over recent years. PLD is usually asymptomatic and not associated with impaired liver function. Symptoms arise mainly from the progressive growth of the multiple cysts or a dominant cyst giving rise to pressure effects on hepatic ducts, vessels and adjacent organs. Most cases require no surgical intervention unless they become symptomatic or complications occur.

A variety of treatments have been recommended. A single large cyst can be obliterated by needle aspiration or instillation of ethanol 98 per cent<sup>1</sup>. However, relapse rates reported from several studies are high and the risks of introducing infection, biliary leakage as well as damage to the normal liver parenchyma adjacent to the cyst remain a possibility. If the contents of the cysts are bile stained, internal drainage into a Roux-en-Y limb of jejunum is advisable. A new surgical procedure described by Lin *et al*<sup>2</sup> which recommends that the superficial cysts be widely fenestrated to allow the deeper cysts to be similarly deroofed, has shown promising results. In cases with symptoms produced by diffuse multiple cystic involvement, partial liver resection combined with extensive fenestration has been suggested<sup>3</sup>.

We report a patient diagnosed with PLD presenting with gross abdominal distension and pain treated by fenestration and segmental liver resection.

### Case Report

Madam YKL is a 56-year-old Chinese lady who first presented in January 1987 with epigastric pain that was sharp and sudden in onset. She was appropriately investigated but defaulted follow-up. Within the same year, she underwent an operation at a private hospital after her symptoms recurred. The nature of the operation is not known. In 1989 she consulted us again with the same complaint. A decision was made to treat her conservatively.

In December 1992, she was readmitted on account of upper abdominal swelling associated with dull pain. However, she did not have fever, vomiting or diarrhoea. She denied being jaundiced and did not have any bleeding tendency or ankle oedema. There was no loss of appetite or weight. There was no known medical illness or any previous history of trauma. She attained menopause four years earlier and was never on oral contraception. There was no history of similar illness in the family.

On clinical examination she was pale but not jaundiced and there was no stigmata of chronic liver disease. The upper abdomen was distended and a right subcostal scar was seen over the right hypochondrium. A large firm nodular mass was felt, measuring 7cm from the right subcostal edge with a span of 22 cm extending from the right to the left hypochondrium. This was not tender, its edges were irregular and there were no ascites. The spleen and kidney were not palpable.

Investigations revealed a mild normochromic, normocytic anaemia with a haemoglobin of 10.7g/dl. Her liver function tests were normal except for slight elevation of alkaline phosphatase (168u/1). The renal function tests were also normal. CT scan revealed multiple cystic lesions involving both lobes of the liver (Figure 1) sparing a small area of normal liver parenchyma seen in segment II. There were no cysts in either kidney.

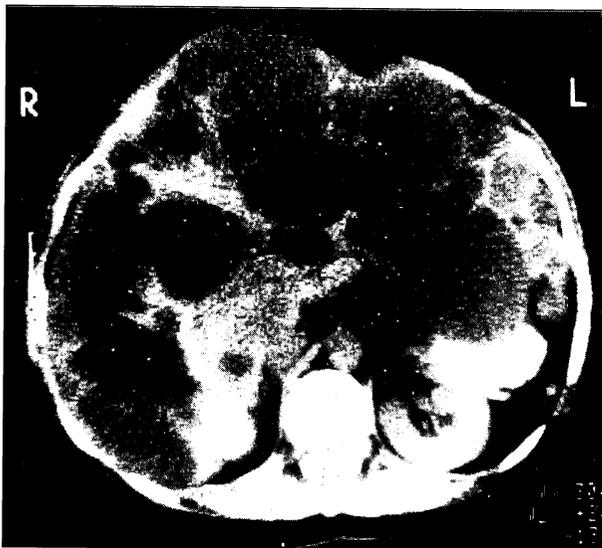


Fig. 1 : CT scan showing multiple cystic lesions of the liver

## CASE REPORT

Laparotomy was undertaken with the intention to reduce the liver bulk by fenestration and if possible segmental liver resection. The right subcostal scar from previous surgery was converted to a roof top incision. As the peritoneum was entered there were intense intra abdominal adhesions posing some technical difficulties. There were multiple cysts of varying sizes seen on the liver surface. These ranged from diffuse multiple small cysts to a few dominant cysts with minimum normal liver parenchyma (Figure 2). There was minimal fluid collection in the peritoneum.

The superficial large cysts were fenestrated first so as to allow access to the deeper cysts. Intraoperative ultrasound was helpful to define the line of resection, keeping away from the hepatic vessels and ducts, and identifying normal liver parenchyma (Figure 3). A small area of normal liver parenchyma was seen in segment II. Segment III of the liver was diffusely involved and this segment was resected. Deroofing and fenestration were performed in two areas of dominant cysts. The cavities created were then patched with omentum. The rest of the laparotomy was normal.

Postoperative recovery was unremarkable and she was later discharged one week after operation. Her symptoms improved and she was well on her first follow-up one month later.



**Fig. 2 : Intra operative findings of multiple cysts on the surface of the liver**



**Fig. 3 : Intra operative use of ultrasound to define the line of resection**

## Discussion

It is generally believed that intrahepatic cysts are derived from the embryological structures called Meyenburg complexes. These complexes are composed of small intralobular bile ducts derived from the developing hepatic parenchyma and their function is to provide a connection between the bile canalicula and the interlobular bile ducts. A maldevelopment during embryogenesis may lead to a persistence aberrant intralobular ducts formation from these complexes, with subsequent intralobular ducts dilatation forming solitary or multiple cysts. The cysts usually contain non bilious fluid as there is no communication between the aberrant intralobular ducts with the biliary tree.

Patients with PLD usually remain asymptomatic, however some slow growing cysts may enlarge with time and produce compression symptoms. Our reviews of literature showed no report on cases of hepatic failure caused by direct liver compression or multiple cyst formation. In most symptomatic cases the patients complain of abdominal distension and chronic abdominal pain. The pain experienced is due to liver enlargement distending Glisson's capsule whereas acute abdominal pain may be due to spontaneous cyst rupture. Occasionally trauma to the enlarged liver may lead to intracystic or

intraoperative haemorrhage. Other less common presentations include jaundice, torsion of a pedunculated cyst, portal hypertension with oesophageal varices or infection presenting as septicaemia, cholangitis or abscesses. The development of neoplastic transformation of adenoma or adenocarcinoma from the Meyenburg complexes is exceptionally rare.

The principle of management of patients with PLD is to control the progression of the larger cyst and to relieve symptoms. In most cases conservative treatment is taken if the symptoms are mild and the patient need to be closely observed. Aspiration may be performed to relieve symptoms or as a temporary measure. Apparently the recurrence rate documented from this procedure is high and it also carries the risk of introducing infection. Aspiration and instillation of alcohol has also been reported<sup>1</sup>. However, the high recurrence rate and the risk of instillation causing destruction to the compressed liver parenchyma does not favour the use of this procedure. The fenestration described by Lin *et al*<sup>2</sup>, involves superficial excision of cysts and deroofting of the deeper cysts through the common wall of the initially excised superficial cysts is able to decompress the enlarged cystic liver, relieve pain and discomfort. This procedure is feasible with the presence of dominant size cysts. In patients with massive cystic liver involvement, complications are frequently encountered as the anatomical plane of the distorted liver is not recognised endangering the vessels, ducts and other adjacent organs. In the presence of multiple small diffuse cysts, partial or segmental liver resection should be considered. Partial liver resection and fenestration described by Armitage and Blumgart<sup>3</sup> appears to be the treatment of choice for symptomatic patients. They suggested resection and fenestration along a projected plane between diffuse cystic areas and spared the normal liver parenchyma by performing serial aspiration to define the planes and the process terminated when normal liver tissue is encountered posteriorly.

We have successfully performed fenestration and segmental liver resection in this patient. Fenestration was performed in two dominant superficial cysts. Subsequently, by deroofting from the common wall of the two dominant cysts, the deeper cysts were accessed. A wider space and area of deroofting were also achieved. The plane of liver resection was well visualised by the used of intraoperative ultrasound (Figure 3). The major hepatic vessels and ducts as well as adjacent structures could therefore be safely defined before resection. The normal liver parenchyma was also safeguarded and the distorted anatomy outlined without much difficulties. The role of intraoperative ultrasound needs to be emphasised.

In conclusion this report illustrates that symptomatic PLD can be safely treated by fenestration and segmental liver resection.

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