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# Caroli's Disease: Functional Diagnosis Using $^{99m}$Tc Diethyl-Iminodiacetic Acid (IDA) Hepatobiliary Scintigraphy

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## ABSTRACT

**A case of Caroli's disease is presented and the findings of various imaging modalities are discussed with emphasis on the role of Technetium $^{99m}$Tc diethyl-iminodiacetic acid scan in reaching the final diagnosis. The 'beaded' appearance of the dilated intrahepatic ducts seen in this scan is believed to be pathognomonic for this condition.**


**Keywords: Caroli's disease, diagnosis, technetium $^{99m}$Tc diethyl-IDA scintigraphy**

## INTRODUCTION

This autosomal recessive condition , first described by Caroli *et al* in 1958, is a congenital segmental cystic dilatation of the intrahepatic biliary ducts representing true ductal ectasia , usually saccular and involve part of or all of the intrahepatic biliary ducts and maintain communication with the remaining biliary system[1]. It is a rare disease that usually presents with recurrent abdominal pain and fever[2].

Early diagnosis and treatment can reduce complications of ascending cholangitis, biliary calculus formation, biliary cirrhosis, portal hypertension, gastrointestinal haemorrhage and septicaemia. Unless the index of suspicion is high the diagnosis can be missed.

Ultrasonography and computed tomography (CT) are useful in detecting the dilated intrahepatic ducts. However, specific diagnosis is not always possible[3] when communication between the intrahepatic cystic lesions and the dilated biliary ducts are not demonstrated.

Cholangiography performed percutaneously or by endoscopic cannulation is the gold standard of diagnosis of this disease. However, this is an invasive procedure and requires special expertise.

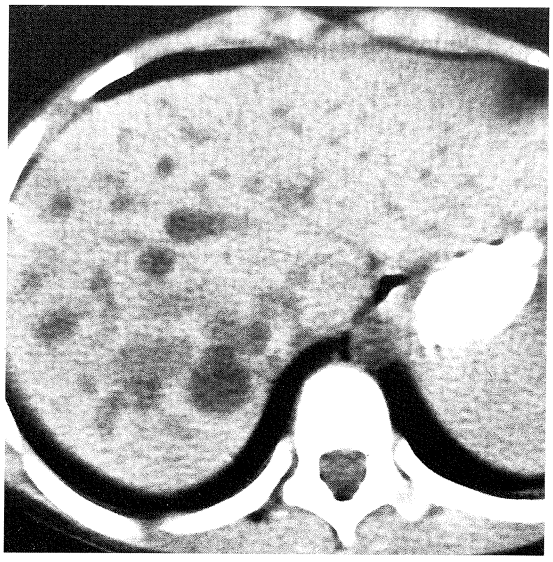
The purpose of this paper is to demonstrate the excellent diagnostic value of $^{99m}$Tc-diethyl IDA hepatobiliary imaging in Caroli's disease when such a situation arises.

## CASE REPORT

An 11-year-old child presented with recurrent febrile episodes for 2 years. These were associated with chills and rigors but no abdominal pain nor jaundice. Physical examination revealed a pale child with a firm palpable liver of 5 cm below the right costar margin. Laboratory investigations revealed a microcytic hypochromic anaemia with sufficient iron stores and a normal electrophoresis. Serum bilirubin and liver enzymes were normal .

An abdominal ultrasound demonstrated hepatomegaly with multiple cystic lesions in both the lobes. The size varies from 2-20 mm. Most of the cysts appeared discrete, although there was a suspicion that some of the cysts may be communicating with each other by dilated tubular structures. There were no detectable intraluminal protrusion nor bridging across the cystic lesions. A CT scan revealed similiar cystic lesions with no central enhancing dots within them after i/v contrast medium injection (Fig 1).

This was followed by a hepatobiliary scintigraphy using $^{99m}$Tc diethyl-IDA which showed uniform uptake of the radiopharmaceutical by the liver parenchyma initially. Subsequent sequential images demonstrated accumulation of radiopharmaceutical in the intrahepatic ducts with multiple foci of increased activities, giving a 'beaded' appearance (Fig 2 and Fig 3). A delayed image showed almost complete clearance of the radiopharmaceutical via the common bile duct into the gut. The gallbaldder was also visualised.



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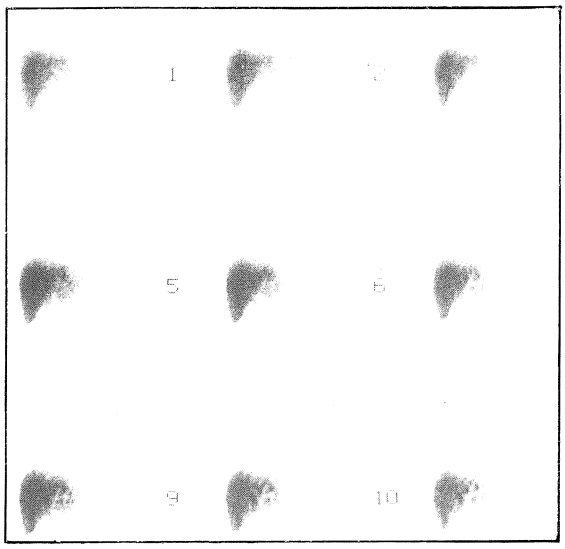

**Fig 2 -** Sequential images following injection of $^{99m}$Tc diethyl-IDA showing initial uniform uptake by the liver parenchyma with subsequent accumulation of the radiopharmaceutical in the dilated intrahepatic ducts.

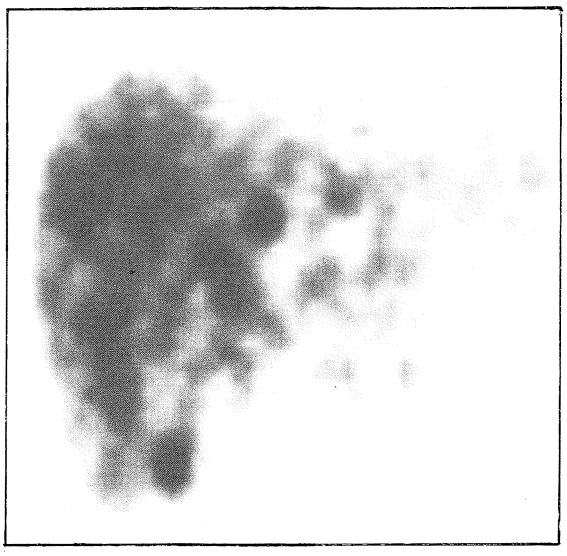

**Fig 3 -** A magnified image demonstrating the dilated intrahepatic ducts with multiple foci of $^{99m}$Tc diethyl-IDA concentration giving a 'beaded' appearance.

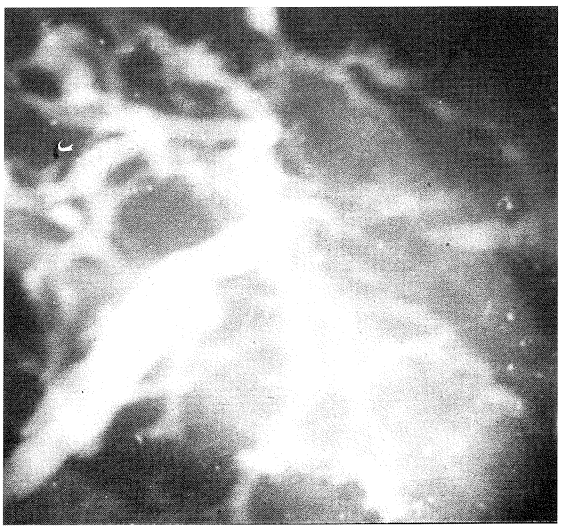

**Fig 4 -** ERCP demonstrating the saccular dilatation of the intrahepatic ducts. The extrahepatic duct appears normal.

Subsequent endoscopic retrograde cholangiography (ERCP) was done and showed entire intrahepatic biliary tree to be abnormal with cystic, saccular dilatation, narrowing and two strictures. The extrahepatic duct was normal (Fig 4). These findings were consistent with Caroli's disease.

## DISCUSSION

In Caroli's disease, the cystic dilatation of the intrahepatic and occasionally the extrahepatic biliary ducts maintain communication with the remaining biliary system[1]. Previously the diagnosis was established by ERCP, PTC (percutaneous transhepatic cholangiography) or surgery. These are however invasive procedures which have their own morbidity and mortality. CT findings were found to be diagnostic when intrahepatic cystic lesions attached to branching low density tubular structures (dilated biliary ducts) were present[4]. The central dot sign[3], which represented portal radicles surrounded by dilated intrahepatic ducts, were found in other cystic disease of the liver besides Caroli's disease[5]. Sonographic findings of dilated bile ducts with vascular radicles described as intraluminar bulbar protrusion and bridge formation were regarded as specific for Caroli's disease[6]. However the CT and ultrasound diagnosis are based on morphological criteria, and where the typical findings are not seen as in this patient, a $^{99m}$Tc diethyl-IDA scan would be the next appropriate investigation. The scintigraphy will establish that the cystic lesions seen on ultrasound and CT scan are due to biliary tract dilatation with communication to the rest of the biliary tract[7]. The 'beaded' appearance which represent dilated biliary ducts[8] which subsequently drain into the normal biliary tract are characteristic if not pathognomonic for this condition.

In addition cystic renal lesions associated with Caroli's disease may be demonstrated as the $^{99m}$Tc diethyl-IDA is excreted via the kidneys[9].

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