

# An 80-Year-Old Lady With Choledocholithiasis In A Cystic Dilated Common Bile Duct: Cause Or Consequence?

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## Abstract

Congenital cysts of the common bile duct (CBD) are rare. Mostly they are found in children and do not typically lead to the formation of common bile duct stones. It is uncertain whether primary CBD stones and acquired choledochal cysts exist. We present an 80-year-old woman presented with epigastric discomfort. Investigation revealed dilatation of the CBD with large concretions. The gallbladder showed no abnormalities and no stones. A cholecystectomy and CBD exploration with stone extraction were performed. Choledochal cysts are usually congenital and discovered in childhood. Congenital CBD cysts are known for their risk of forming a bile duct malignancy with an increasing incidence with age. Presentation at the age of 80 years has not been reported before. We believe that in this patient the underlying aetiology was a choledochal cyst-like dilatation acquired during life leading to stasis of bile and formation of primary CBD stones.

## INTRODUCTION

Cholelithiasis is a very common disease in the western world. In the Netherlands annually 20.000 patients are admitted for symptomatic gallstones and 17.000 cholecystectomies are performed<sup>1</sup>. Common bile duct (CBD) stones usually occur in association with cholecystolithiasis. It is generally accepted that the majority of stones found in the CBD are secondary stones having migrated from their primary site of origin in the gallbladder into the CBD through a patent cystic duct or a cholecysto-biliary fistula<sup>2,3,4</sup>. CBD stones are occasionally present in patients with an apparently normal gallbladder without stones<sup>5,6,7</sup>. These primary CBD stones may have formed de novo within the CBD as a result of ampullary stenosis, periampullary diverticula, cholangiocarcinoma, haemolytic anaemia or choledochal cysts. Congenital cysts of the CBD are rare, occurring in 1:100,000-150,000 live births and are typically found in children<sup>8</sup>. Here we report a case of an 80-year-old patient combining a cystic CBD with primary CBD stones.

## CASE REPORT

An 80-year-old unjaundiced Caucasian woman was seen in the outpatient clinic with a 3 weeks history of epigastric discomfort. There was no history of biliary colic, jaundice, fever or chills. Serum bilirubin (18,5  $\mu\text{mol/l}$ ) and gamma-glutamyl transferase (58  $\text{u/l}$ ) were slightly elevated (normal 2-17  $\mu\text{mol/l}$  and 2-50  $\text{u/l}$  resp). Hemoglobin and

serum amylase were normal. Ultrasonography revealed marked dilatation of the CBD with large concretions, but no abnormalities of the gallbladder or pancreato-biliary junction. This was confirmed by endoscopic retrograde cholangiopancreatography (ERCP) (figure 1).

Figure 1



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An attempt at extracting the CBD stones after adequate endoscopic sphincterotomy (ES) was not successful. A stent was placed for decompression. During laparotomy a cyst-like dilated CBD was encountered with a normal gallbladder and cystic duct. There were no signs of malignancy of the biliary tree. A cholecystectomy and CBD exploration was performed. Several brown friable stones were extracted, the largest being two cm in diameter (figure 2).

**Figure 2**



A choledochoscopy was performed and apart from dilatation of the CBD no abnormalities were found. Because of patient's high age and assumed aetiology of the dilatation, treatment was limited to stone extraction. A T-tube was left in place. Postoperatively the patient recovered well and serum liver enzyme tests and amylase remained within the normal range. A T-tube cholangiogram ten days after surgery showed persistent cystic dilatation of the CBD but no remaining stones and a rapid flow of contrast medium to the duodenum (figure 3).

**Figure 3**



The pathology report of the gallbladder revealed no abnormalities and no stones. The patient was readmitted 18 days following discharge with recurrent abdominal pain and elevated serum liver enzymes. Repeat ERCP showed a very wide-open papilla of Vater after adequate ES and persistent dilatation of the CBD without concretions. The patient was diagnosed as having ascending cholangitis and recovered well after treatment with antibiotics.

### DISCUSSION

The presented case of an 80-year-old patient with a cystic CBD in the absence of a biliary tree malignancy combined with CBD stones is interesting because it illustrates several aspects of the aetiology of CBD stones and CBD dilatation. We describe the case of the oldest patient presenting with a choledochal cyst in the recent literature. In a group of 1,433 patients described by Yamaguchi et al. the ages ranged from 6 days to 78 years<sup>8</sup>. The mild nature of our patient's complaints and the very mild abnormalities of the laboratory findings are not exceptional both in patients with CBD dilatation and in patients with CBD stones<sup>3</sup>. Choledochal cysts are closely related to the formation of primary CBD stones<sup>6</sup>. Isolated fusiform dilations of the extrahepatic biliary tree, type I cystic lesions according to Todani, account for the majority of all choledochal cysts<sup>8,9</sup>. The classical clinical triad in choledochal cysts of abdominal pain, jaundice and a right hypochondrial mass has been frequently reported in children. A complete triad in adults however is rare<sup>10,11</sup>.

In a group of 88 patients with adult-type cystic dilatation of

the CBD, reported by Matsumoto et al. 70 patients had gallstones.<sup>6</sup> In 48 of these 70 patients there were only stones in the CBD and not in the gallbladder. The incidence of bile duct malignancy in patients with congenital choledochal cysts increases with age from 14% for those older than 20 years to 50% for patients aged over 50.<sup>12,13</sup> For this reason complete choledochal cyst excision is advocated.<sup>14</sup> In view of the rise of the incidence of malignancy with age, the absence of malignancy in this patient despite patients' extraordinary advanced age, strongly supports the theory that although most choledochal cysts are congenital, cystic dilation of the CBD can develop during life.<sup>15</sup> The pathogenesis of acquired cystic enlargement is unknown. A relative distal obstruction of the CBD caused by a pancreatobiliary junction anomaly subsequently leading to chronic ascending inflammation of the biliary tree and weakening of the CBD wall is believed to be important in the pathogenesis of CBD dilatation.<sup>16,17,18</sup> In some parts of the world biliary tract obstruction is often the result of an infection by parasites such as *Ascaris lumbricoides* or *Fasciola hepatica*.<sup>19</sup> Our patient however, did not visit or reside in parts of the world where these parasites are endemic. Because dilatation of the CBD is also seen in patients without obstruction it has been suggested that this dilatation represents an entity similar to achalasia of the esophagus. In most patients however the wall of the CBD does not contain smooth muscle.<sup>20</sup>

Alternatively, in our patient choledocholithiasis may have caused dilatation of the CBD. Although septic cholangitis is the most dramatic manifestation of choledocholithiasis, a more gradual, progressive obstruction may also occur, with minimal symptomatology. Charcot's triad (jaundice, pain and fever) is present in less than 25% of patients.<sup>21</sup> In our patient the gallbladder was thin-walled, without stones or inflammatory signs and the cystic duct was of normal size. Therefore it seems reasonable to assume that our patient had primary CBD stones. In the presence of primary common bile duct stones, it is not uncommon that the gallbladder does not contain stones. Primary CBD stones can form de novo in the CBD as a result of stasis or infection.<sup>22,23</sup> In recurrent cholangitis bacterial deconjugation of bilirubin leads to the formation of insoluble bilirubin salts and thus to primary biliary stones.<sup>24,25,26</sup> Madden described that 47% of the patients in his series with primary stones had ampullary stenosis.<sup>27</sup> In our patient, there was no evidence of recurrent infection or ampullary stenosis contributing to the formation of primary CBD stones.

Theoretically it is still possible that despite the advanced age of the patient, a congenital choledochal cyst-like dilatation of the CBD was present. On the basis of the considerations mentioned above however, especially regarding patients' advanced age and absence of stones in the gallbladder, we conclude that in our patient a weakening of the CBD wall resulted in a cystic dilatation, subsequently leading to stasis of bile and formation of primary CBD stones.

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