Congenital Type 1C Choledochal Cyst: Clinical Presentation and Surgical Treatment

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Abstract

**Introduction,** Choledochal cysts are rare anomalies of intra and extrahepatic biliary ducts in children and adults. In all cysts, a complete excision is recommended to prevent chronic inflammation and malignant transformation.

**Presentation of Case,** An 8-year old female patient admitted to the paediatric ward as a case of acute pancreatitis. The patient complained of severe upper abdominal pain of three days duration radiating to inter-scapular area associated with vomiting, mild itching and fever. A past history of recurrent attacks of mild abdominal pain and vomiting of two years duration was elicited. Clinical examination revealed epigastric and upper right abdominal tenderness, mild ting of jaundice with no hepatomegaly.

**Discussion,** A diagnosis of a congenital type 1C choledochal cyst was achieved following resonance cholangio-pancreatography (MRCP) examination. During laparotomy, the choledochal cyst extended from the common hepatic duct just below the junction of right and left hepatic ducts down into the bile duct as far as the point proximal to its junction with the main pancreatic duct. Resection of the whole segment of choledochal cyst together with the gall bladder, assisted Roux-en-Y hepatojunostomy was performed and the abdominal wound was closed leaving no drain. Postoperatively, the patient unremarkably recovered, the abdominal pain and jaundice subsided and was discharged on the 5th postoperative day.

**Conclusions,** the procedure of laparatomy fine dissection and resection of choledochal cyst and gall bladder with Roux-en-Y hepatojunostomy is still superior to endoscopic approach since it is grossly performed with less chance of postoperative complication
Introduction:

Choledochal cysts are rare developmental anomalies involving either extrahepatic or intrahepatic biliary ducts, or both\textsuperscript{1-3}. Epidemiologic studies on choledochal cysts shows higher frequency in females than in males (3:1 to 4:1) with remarkable high incidence of 1 in 1000 births among Asian population compared to incidence of 1 in 100,000-150,000 in Western countries \textsuperscript{4, 5}. About two-thirds of diagnosed cases of choledochal cysts in Asia occur in Japan \textsuperscript{6}.

Vater \textsuperscript{7} was first to provide an anatomical description of the choledochal cyst. Alonso-Lej et al \textsuperscript{8}, based on analysis of 96 reported cases of choledochal cysts have devised 3 categories classification and their therapeutic strategies. However, Todani et al \textsuperscript{9} has refined this classification system to include 5 categories which become the most acceptable and applicable by most authors.

The aetiology of these cysts remains under speculation, as many theories and explanations have been proposed \textsuperscript{1}, however the Babbitt’s theory \textsuperscript{10} of choledochal cysts formation remains the most popular and accepted. This theory attributes the cyst formation to abnormal pancreaticobiliary duct junction (APBDJ) outside the ampulla of Vater resulting in a long common channel between the bile duct and main pancreatic duct allowing the reflux of pancreatic juice into the bile duct. This reflux results in activation of pancreatic enzymes causing inflammation and weakness of bile duct wall leading to cyst formation.

The present article outline a case of congenital choledochal cyst involving large segment of bile and common hepatic ducts presented with severe abdominal pain, fever and mild jaundice. The applied surgical procedure is described in detail and is compared with other surgical techniques endorsed in the treatment of these congenital choledochal cysts.
Case report & surgical procedure

An 8-year old female patient with history of repeated attack of upper abdominal pain and bouts of vomiting for the last two years was admitted to the emergency unit of Prince Hamza Hospital, Amman, Jordan during February 2011 complaining of sever upper abdominal pain radiating posteriorly to interscapular area with attacks of vomiting, yellow discoloration of skin, mild itching, and dark colour urine.

On clinical examination she looked sick, cachexic, and febrile with forward bending posture at the trunk. Mild ting of yellow discoloration of skin and conjunctiva was noticed, and on palpation, sever tenderness in the epigastrium and right hypochondrium was elicited with non-palpable liver. She was admitted to the paediatrics unit as a case of acute pancreatitis.

Laboratory investigations revealed normal serum amylase level, elevated serum levels of direct bilirubin, alkaline phosphatase, ALT and AST liver enzymes (data not shown).

Magnetic resonance cholangiopancreaticography (MRCP) revealed 10 x 7 cm fusiform cystic swelling of the common bile duct extending into the common hepatic duct with normal size gall bladder and cystic duct (Figs. 1 and 2). The intrahepatic biliary ducts were slightly dilated, and a diagnosis of Type 1C choledochal cyst was concluded. The patient was kept on IV fluid, antibiotics and analgesic waiting for surgical interference.

Laparatomy with upper right transverse incision was performed and a large fusiform cystic swelling involving the common bile duct and extending into the common hepatic duct up to a level above the opening of cystic duct. The gall bladder, cystic duct and the liver appeared normal. The gall bladder bed was dissected and freed after ligation of cystic vessels to assist dissection and freeing the cyst from portal vein and hepatic artery. The cystic swelling was dissected from porta hepatis down to retrodudenal part of bile duct. The cystic bile duct was ligated proximal to the junction with the main pancreatic duct, excised and reflected up toward the porta hepatis. The whole segment of cystic bile duct, common hepatic duct, cystic
duct and gall bladder was excised. The jejunum 40 cm below the ligament of Trietz was transected and the ostium of distal segment was closed using Vicryl 4/0 continuous sutures. The closed-end distal segment of jejunum was lifted to porta hepatis and sutured with the ostium of the remaining part of common hepatic duct (hepaticojejunostomy) by end-to-side interrupted full suture using Vicryl 4/0 (Fig 3). The ostium of the proximal cut segment of jejunum was sutured end-to-side with distal segment of jejunum (Roux-en-Y procedure) using Vicryl 4/0 interrupted single layer seromuscular sutures (Fig 3.). The abdominal incision was closed without drain, and the excised specimen was sent for histopathology. The histopathologic examination of the excised tissue confirmed the diagnosis of benign choledochal cyst involving common bile and hepatic ducts with normal gall bladder tissue (Fig. 4).
Postoperatively, the patient recovered unremarkably, the pain and jaundice disappeared and was discharged on the 5th postoperative day.
Discussion

Choledochal cysts are congenital anomalies of biliary tract being more common among Asian population [4-6]. Developmentally, the biliary ducts develop from a hepatic diverticulum in the ventral wall of caudal end of primitive foregut early in the 4th week of intrauterine [11, 12]. This small diverticulum will develop into extrahepatic and intrahepatic biliary ducts, gallbladder, and ventral pancreas. Recanalization of the lumen of common duct starts at the end of 5th week and moves slowly distally. By the 6th week, the common duct and ventral pancreatic bud rotate 180 degrees clockwise around the duodenum. Early in the 7th week, the bile and pancreatic ducts end in closed cavities of the duodenum. The muscle of the sphincter of Oddi develops from a concentric ring of mesenchyme surrounding the preampullary portion of bile and pancreatic ducts.

The aetiology of choledochal cysts remains speculative, however the pathogenesis most probably is multifactorial. In majority of patients with choledochal cysts, a developmental abnormality at the junction between the common bile duct and the main pancreatic duct can be demonstrated. This occurs when the pancreatic duct empties into the common bile duct more than 1 cm proximal to the ampulla of Vater [10]. Miyano and Yamataka [6] have documented such abnormal junction in 90-100% of patients with choledochal cysts. They attributed the formation of choledochal cysts to the reflux of activated pancreatic proenzymes into the common bile duct which cause damage and weakness of the bile duct wall. Defects in the epithelialization and recanalization of developing bile ducts, congenital weakness of the duct wall due to distal aganglionosis have also been implicated [13, 14].

The choledochal cyst have been classified according to the system published by Todani et al. [9] into five major classes;
1- Type I cysts being the most common and represent about 80-90%. They consist of saccular or fusiform dilatations of the common bile duct, which involve either a segment of the duct or the entire duct. This type is further subdivided into; type IA is saccular in shape and involves either the entire common bile duct or the majority of it; type IB is saccular and involves a limited segment of the bile duct; and type IC is more fusiform in shape and involves most or all of the bile and common hepatic ducts.

2- Type II choledochal cysts appear as an isolated diverticulum protruding from the wall of the common bile duct with or without involvement of common hepatic duct.

3- Type III choledochal cysts arising from the intraduodenal portion of the common bile duct and alternately termed as choledochocele.

4- Type IV cysts are multiple saccular dilatations of either intrahepatic or extrahepatic bile ducts, or both. They are subdivided into; Type IVA consist of multiple dilatations of the intrahepatic and extrahepatic bile ducts type; and type IVB choledochal cysts which are multiple dilatations involving only the extrahepatic bile ducts.

5- Type V or Caroli disease consists of multiple dilatations limited to the intrahepatic bile ducts.

In our present case, the excised cyst represented the category of Type 1C choledochal cyst.

Shah et al [15] in a retrospective study on 32 children and 47 adults with choledochal cysts in regard to their presentation, management, histopathology, and outcomes, concluded that; a history of biliary surgery, pancreatitis, cholangitis, early postoperative complications, and late postoperative complications occurred more frequently in adults than in children; the classic triad of abdominal pain, jaundice, and palpable right upper abdominal mass were more frequent in children than in adults; fibrosis of the cyst wall was peculiar to children; and the signs of inflammation, hyperplasia, and long-term complications were more frequent in
adults. From these findings, they concluded that choledochal cysts of children and those of adults should be considered as separate entities.

Several surgical procedures have been employed in the treatment of choledochal cyst [16-19]. These procedures vary between laparotomy and laparoscopic approaches, however laparoscopic complete cyst resection, assisted Roux-en-Y reconstruction and hepaticojejunostomy remains the procedure of choice by majority of surgeons.

In conclusion, we aimed to resect extrahepatic biliary ducts as much as possible preserving the last segment of bile duct joining the main pancreatic duct. The procedure applied reduces the risk of subsequent malignant changes in the remaining stump. Others have advocated a procedure leaving a longer stump of common hepatic duct with mucosal resection as alternative measure of protection against cancerous changes. Our personal experience and view suggest that a longer de-mucosal stump is more liable to malignant changes, and stump stricture is quiet likely to occur.
Conclusion

Developmental choledochal cysts are clinically presented during childhood or in adult life, and their risk is being complicated by inflammation, rupture and peritonitis, and malignancy. The present report illustrates a case of type 1C choledochal cyst in an eight years old female which was hospitalized as a case of acute pancreatitis. The diagnosis was approved by MRCP and by an upper right transverse laparotomy, the cyst was completely excised and Roux-en-Y hepatojejunostomy was performed.
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References


**Consent:** A written informed consent was obtained from the patient for publication of this case report and accompanying images.

**Conflict of Interest:** All authors have no conflict of interest
Legends of Figures:

Figure 1 (a and b): MRI pictures of choledocal cyst in a 10-years old female prior to surgical excision. The cyst is fusiform-shaped (type 1C) involving the bile and common hepatic ducts.

ChC=choledochal cyst; CHD=common hepatic duct; RHD=right hepatic duct; LHD=left hepatic duct; DU=duodenum; CBD=common bile duct.

Figure 2. Cross sectional MRI picture of the choledochal cyst. ChC= choledochal cyst; GB=gall bladder; RK=right kidney; LK=left kidney.

Figure 3. Schematic drawing of the applied Roux-en-Y hepatoenterostomy surgical procedure.

Figure 4. Light microscope picture of the excised choledochal cyst (a) showing fibro-muscular tissue partially lined by simple epithelial mucosa with no malignant changes. The gall bladder tissue (b) appeared normal. (a and b x10)