

Choledochal Cyst: A Report of 3 Cases and a Review of its Treatment and Outcome

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Abstract

Choledochal cyst (CC) is not common in Africans. Unlike biliary atresia, this condition commonly presents in late infancy and early childhood, and occasionally also in adults. Typical pattern of presentation is with abdominal mass, abdominal pain and seldom intermittent jaundice. These conventional presentations can be sometimes heralded by cholangitis, malnutrition, and failure to thrive in some category of patients with delayed presentation.

Case 1 was a 2-month-old girl who developed fluctuating jaundice on the 5th day of life, associated steatorrhea, and yellowish discoloration of the urine and occasional bilious vomiting. Ten days before presentation she developed low grade fever, excessive crying and refused to suck. She was resuscitated and hepaticoduodenostomy was performed between the second part of the duodenum and the stump of the common hepatic duct.

Case 2 was a 4-year-old girl who presented with a 5-month history of fluctuating jaundice associated abdominal pain, epigastric swelling, weight loss and occasional non-bilious vomiting. A retrocolic end-to-side Roux-en-Y hepaticoduodenostomy was performed after nutritional rehabilitation.

Case 3 was a 5-year old girl with intermittent jaundice noticed 2-months before presentation, right hypochondrial swelling and colicky right sided abdominal pain almost of same duration. Intraoperatively features of likely cirrhotic liver changes in addition to a huge choledochal cyst were seen. An end to side Roux-en-Y hepaticoduodenostomy was established to the stump of the common hepatic duct.

All the cases did well after surgery and were discharged. Regardless of the clinical features, and the morphological changes in the liver, excision of the cyst with establishment of free flowing hepatoenterostomy should be done. Patients' general outlook can be immediately improved after excision of the cyst with hepatoenterostomy. Because the complications of caused by bile flow obstruction are remarkably reduced or rather eliminated. We hereby report three cases we recently managed in our centre with complications of choledochal cyst which improved after excision of the cyst with creation of hepatoenterostomy.

Keywords: Choledochal cyst; Hepaticoduodenostomy; Hepaticoduodenostomy

Introduction

Choledochal cyst (CC), also called 'bile duct cyst' is an uncommon congenital condition and one of the rarest causes of obstructive jaundice in infancy; characterised by simple, multiple extra or intra hepatic biliary cystic dilatations that can occur on any part of the bile duct. It has significant geographical variation, rarely seen in Blacks as in Caucasians and Asians [1]. Incidence as high as 1 per 1000 in Asia (Japan), and as low as 1 per 100 000-150 000 in the West has been reported [2,3]. Seventy to eighty percent (70-80%) of cases occur in infants and children, and the remaining 15-20% occur in adults with a male to female ratio of 1:4 [4,5]. Few cases have been reported in Nigeria and Africa [6-9]. This is a report of 3 patients, including a review of the pathogenesis, current treatment and outcome.

Case 1

A 2-month-old girl developed fluctuating jaundice on the 5th day of life with steatorrhea, yellowish discoloration of the urine and occasional bilious vomiting. Ten days before presentation she developed low grade fever, excessive crying and refused to suck. Her temperature was 38.3°C; she was dehydrated and jaundiced, with a respiratory rate of 30 cycles/min, heart rate-160 beats/min, and weighed 4.9 kg (which is 76% of the expected weight for her age). The abdomen was uniformly distended; a non-tender cystic mass was palpable in the right hypochondrium which moved with respiration. The liver span was normal. Abdominal ultrasound scan (Figure 1) showed a huge intra-abdominal cystic lesion arising in the region of the liver and extending from the epigastrium to the pelvis, more to the right and measures about 10.87cm × 9.60 cm × 8.86 cm.



Figure 1: Ultrasound finding of a cystic choledochal cyst compressing on the liver.

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The patient was resuscitated with intravenous fluids; broad spectrum before surgery. Intra operatively, a bile-containing cystic mass consistent with Todani's [10,11] type Ia choledochal cyst arising from the common bile duct (CBD) extending proximally beyond the cystic duct and distally toward the duodenum was found. The liver and gall bladder were grossly normal. The cyst was completely resected along with the gall bladder and a hepaticoduodenostomy was performed between the second part of the duodenum and the stump of the common hepatic duct. The peritoneum was closed over a sub hepatic drain, which was removed on the 5th postoperative day and discharged on the 10th day after surgery. The histology was of a partly denuded cyst wall, partly covered with columnar epithelium, with intense transmural acute inflammation and congested vascular channels consistent with choledochal cyst. The patient was well at 6-months of follow up before defaulting from further follow up visits.

Case 2

A 4-year-old girl presented with a 5-month history of fluctuating jaundice associated abdominal pain of 3-months' duration, epigastric swelling, weight loss and occasional non-bilious vomiting. The stool was never pale. On examination she was chronically ill looking, febrile with axillary temperature of 38.9°C, pale, icteric, and dehydrated. The weight was 6.5 kg (54% of the expected weight for her age). The epigastrium was distended by a firm tender non-mobile mass with a smooth surface and well-defined lower edge. Abdomino-pelvic ultrasound scan revealed a huge well-circumscribed intra-abdominal cystic lesion with coarse internal echoes within it, located postero-inferior to the liver and extending from anterior abdominal wall to the spine, measuring about 10.03 cm × 9.26 cm × 6.98 cm. Total bilirubin ↑30 mmol/L (ref value 1.7-17.1 mmol/L), conjugated bilirubin ↑21 mmol/L (ref value 1.7-8.5 mmol/L), alkaline phosphatase 105 iu/L (ref value 60-170 iu/L), total protein 42g/l (ref value 58-80 g/L), and albumin 27 g/L (ref value 35-50 g/L). The patient was nutritionally rehabilitated, resuscitated and prepared for laparotomy. Intraoperative findings (Figure 2) were: a type Ia CC covered by a patch of an oedematous omentum on its anterior and lower parts, and a collapsed gall bladder. The cyst was mobilised, but completely excised in peace-meal from the portal structures, to reduce risk of injury to the portal structures because of the oedema. The gall bladder was also resected. The common hepatic duct and the intra-hepatic ducts probed and lavaged with saline to rid the ductal system of sludge, a Roux-en-Y end-to-side retrocolic hepaticojejunostomy was performed to about 35 cm isolated limb of the jejunum. Total serum and conjugated bilirubin stabilised on the first postoperative week. Histology showed a fibrocollagenous cyst wall covered by columnar epithelium with moderate cytoplasm, focal areas of metaplasia, intense

transmural acute inflammation and congested vascular channels consistent with choledochal cyst, no evidence of malignancy. The patient was discharged on the 2nd week postoperatively. She was follow up just for 6-weeks before defaulting subsequent follow up visits.

Case 3

A 5-year old girl with intermittent jaundiced noticed 2 months before presentation, and right hypochondrial swelling, colicky right side abdominal pain of almost same duration. Sometimes the pain gets relief with analgesics (paracetamol). She also has intermittent low grade fever, acholic stool and dark coloured urine. The patient had been oral herbal drugs for 2-weeks prior to presentation. On general examination she was icteric, afebrile, and weighed 13.5 kg which is 75% of the expected weight for age. The right hypochondrial mass firm, nontender, immobile, extending towards the epigastrium laterally and the right iliac region inferiorly. The liver was enlarged, 6 cm below the costal margin with blunt edges. No demonstrable ascites. Ultra sound scan of the abdomen revealed a well-defined cystic mass in the right hypochondrium measuring 7.8 × 7.8 × 6.8 cm, with dilated intra and extrahepatic biliary ducts, dilated gall bladder, and hepatomegally of 18 cm. The urinalysis was positive for bilirubin. Total and conjugated bilirubins were elevated, total bilirubin 66.7 umol/L (ref value 1.7-17.1), conjugated bilirubin 58 umol/L (ref value 1.7-8.5). The PT 12 seconds (range 10-14 seconds), PTTK 28 seconds (range 25-35 seconds), and the INR 1.0. The serum biochemistry, hematological parameters and viral markers were within normal limits. Two-pints of fresh whole blood were made available before surgery. Intraoperatively a type Ia choledochal cyst with pericyclic adhesions, dilated common hepatic, dilated right and left extra hepatic ducts, enlarged dusky looking liver with features of cirrhotic changes (Figure 3). The cyst completely submucosally dissected and an end-to-side Roux-en-Y hepaticojejunostomy was established to the stump of the common hepatic duct. The patient had recurrent episodes of fever for about 2-weeks which was managed antibiotics (ceftriaxone and metronidazole). She was discharged 5-weeks after surgery. The patient was well at her last follow up 3-months ago. Recent liver ultrasound scan at her last follow showed reduction in size of her liver from 18 cm preoperatively to 13 cm.

Discussion

Pathogenesis

Though biliary duct cyst dilatations are purely congenital, some theories have tried to explain its pathogenesis. The long common channel theory popularised by Babbit in 1969 based on the concept of abnormality in the union of the pancreatico-biliary duct is one of these



Figure 2: Intra-operative finding of the cyst collapsed gall bladder.



Figure 3: a) Choledochal cyst with features of cirrhotic liver changes, b) Cyst wall after excision.

theories [12]. It states that children with abnormal pancreaticobiliary duct junction (APBDJ) are predisposed to high serum level of pancreatic amylase in the gall bladder from reflux of pancreatic juice into the common bile duct leading to post-inflammatory fibrosis and weakness in the wall of the bile ducts in patients with long common channel greater than 10 mm. A higher pressure gradient generated in the pancreatic duct than in the common bile duct in APBDJ predisposes to reflux of pancreatic juice rich in trypsin into the bile duct that can aid in the inflammatory process. Further, more the epithelial lining of the APBDJ can secrete enterokinase which can also indirectly aid in the post-inflammatory weakness of the biliary wall through the production of phospholipase A2, a potent proinflammatory mediator [13]. The long common channel theory is rational, it along is weak in explaining the development how choledochal cyst occurs [14]. Because some population of patients have been found with APBDJ and with high levels of pancreatic reflux into the gall bladder, and yet without cystic dilatation of the common bile duct. Studies found that the chemical content of the pancreatic fluid is not strong enough to elicit significant inflammatory reaction as to cause choledochal cyst in puppies [15]. Hence therefore, the post-inflammatory weakness dilatation in long common channel APBDJ, likely develop in the presence of a distal obstruction in the common bile duct [16,17]. The distal common bile duct obstruction could be due to congenital distal bile duct obstruction, stone formation, scarring in the distal common bile duct, sphincter of Oddi motility dysfunction (as seen in Hirschsprung's aganglionosis due to paucity of ganglion cells), [18] persistence of epithelial membrane or over production of epithelial cells during embryologic development or abnormality of the valve at the ampulla of Vater. In three patients we managed the cysts narrowed out distally toward the duodenal portion, which fit into the concept of a probable congenital distal obstruction to the cyst.

Some authors are of the opinion that clusters of all the anatomical types of choledochal cyst can be explained by the common channel theory, except types I and IV choledochal cyst [19,20]. The suggested pathogenesis of the remaining types are: i) type II CC could be due to an end stage of a healed prenatal rupture of the wall of the common bile duct, because the type II is just a simple diverticulum on the common bile duct with a narrow or wide stalk; ii) type III which invariably should be called (cholechocele), because it could be due to simple congenital duplication of the duodenum or may be just a variant of duodenal duplication cyst as duodenal or biliary epithelium have been found lining the cyst wall [21]; iii) type V also called (Caroli disease) has been linked to ductal plate malformation in which faulty remodelling, and selective resorption of the ductal plate aid in its development [22]. Hence, types II, III and V has no aetiologic relationship to the common channel theory. It is therefore rational that because of the differences in aetiopathogenesis of choledochal cyst an anatomical classification should be more appropriately fit for each of the numerical types described by Todani [23]. Therefore, types I is choledochal cyst, type II is choledochal diverticulum, type III is cholechocele, type IV is a bile duct cyst and type V is Caroli's disease.

Classification

For ease of understanding the classification, the term biliary duct cystic dilatation should preferably be used for the clusters of all the anatomical types of the cyst. Instead the term choledochal cyst should be reserved only to cystic dilatations arising on the choledochus (the duct length between the common hepatic duct and the ampulla of Vater), whereas the term bile duct cyst should be used for those cystic dilatations arising from the right and left extra hepatic ducts and intra hepatic ducts.

The original anatomical classification of choledochal cyst are as follows: type I: (Ia) cystic dilatation of the whole CBD, (Ib) is either a localised, focal or segmental dilatation of the entire common bile duct (CBD), and (Ic) is a fusiform dilatation of the entire common bile duct which may extend to the pancreatico-biliary duct junction (PBDJ). Type Ic is difficult to be differentiated from another form of cystic dilatation of the choledochus called 'Form frusta' [24] (not a fully developed form of CC) which has been described as a condition with similar clinical and histopathological features with CC. Children with it also do have abnormal pancreatico-biliary duct junction (APBDJ), obstructive jaundice and abdominal pain in the absence of an obvious biliary duct dilatation. Surgical excision is therefore pertinent in order to relieve symptoms and reduce the risk of complications. Type II is a diverticulum on the CBD which could be anywhere along CBD; type III also called cholechocele (because its morphology has similar to ureterocele), is an intra-duodenal dilatation of the CBD. Type IV preferably called bile duct cyst, because it primarily involves either the intra or extra hepatic ducts. It is subdivided into two: IVa is multiple intra hepatic and extra hepatic cystic duct dilatations; IVb are multiple extra hepatic cystic duct dilatations. The type IVb bile duct cyst looks like a "bunch of grapes" or "string of beads" based on its appearance on radiology [25]. Type V bile duct cyst is a multiple communicating intra hepatic cavernous ectasia, also called Caroli's disease [26]. This entity should be distinguished from Caroli's syndrome which is just an isolated cystic intra-hepatic biliary dilatations, whereas Caroli's disease is has characteristic cystic intra-hepatic duct dilatations and congenital hepatic fibrosis [27].

Diagnostic evaluation

Type I choledochal cyst is so far the most common type seen in about 70-90% of cases as we have seen in all the cases we managed, followed by type IV (10-35%), type II (2-5%), type III (1.4-4%) and type V (1%) [28,29].

Generally, 80-90% of choledochal cyst present in the infancy and childhood, [30] the remaining 20% of cases are seen in adults. The less frequent ones are the types that usually present asymptomatic [31,32]. They include choledochal diverticulum, cholechocele and the Caroli's disease. Types I and IV choledochal cysts tend to present relatively early, because of the associated bile duct obstruction either from inspissated bile stone, bile plugs, pancreatic stones, congenital distal bile duct stenosis which can easily predispose to obstructive symptoms in types I and IV choledochal cysts [33,34]. All the cases we managed had distal narrowing associated with the cyst which could probably be congenital distal bile stenosis. One patient had biliary sludge in the common hepatic duct that we had to irrigate the hepatic ducts with saline after excision of the cyst.

The clinical presentation is more of abdominal mass and jaundice in the infants, and more of abdominal mass, jaundice and abdominal pain in older children [35]. The only infant in our case had the complaint of abdominal mass and jaundice, whereas the other two children had the classical triad of abdominal mass, intermittent jaundice and abdominal pain commonly seen in older children with choledochal cyst. Jaundice in choledochal cyst is often intermittent because the obstruction is rarely complete unlike in biliary atresia. Character of jaundice is an important ominous symptom in differentiating obstructive jaundice due to biliary duct cystic dilatation from obstructive jaundice due to biliary atresia.

There were other constitutional symptoms in our patients which could be due to cholangitis, as fever was present in all of them. All the

patients were under weight, which could not be unconnected with the complications of the cyst. Cholangitis, pancreatitis, choledolithiasis, pancreatic calculi, and sometimes metaplastic changes are common complications of choledochal cyst. Especially cholangitis is commonly associated with types I and IV choledochal cyst because of recurrent reflux of pancreatic juice in the presence of APBDJ [36,37]. Types I and IV choledochal cyst are also the most frequently seen, thus report about its incidence of complications are frequently documented. The risk of complications increased with age at diagnosis of choledochal cyst, because the duration of stasis, and chronic inflammation, liver injury is increased with late diagnosis [38]. One of the cases we managed had started showing features of cirrhotic changes in the liver, which could be due to chronic inflammatory injury to the liver. The most dreaded of complications of the choledochal cyst is cancer. It is rarely symptomatic except when advanced, occurs 10-15 years earlier in patients with choledochal cyst than in the general population [39,40]. About 40-50% occur in the gall bladder, and 5-10% in the bile duct [41]. Adenocarcinoma 73-84%, followed by anaplastic carcinoma 10%, undifferentiated cancer 5-7%, squamous cell carcinoma 5% and others 1.5% were different histological types of malignancies arising from the cyst described [42-44].

Apart from ultrasound scan which can detect 90% of CCs in good hands, endoscopic retrograde cholangiopancreatography (ERCP) can detect coexisting APBDJ and other associated anomalies with CCs [45]. Incidence of associated anomalies in CCs have not been determined but isolated cases of pancreatic aplasia, congenital absence of the portal vein, heterotopic pancreatic tissue, duodenal atresia, cardiac lesions have been reported [46,47]. Our patients can only afford ultrasound scan, MRI though available but it has very significant financial constraints on the patients and was not requested. Magnetic resonance cholangiopancreatography (MRCP) can replace ERCP in patients with pancreatitis for anatomical diagnosis of the biliary tree and other associated anomalies.

Intra operative cholangiography will be necessary if the preoperative information was not sufficient. However, it could be difficult if the cyst is very large. But, it is valuable in characterizing the anatomy of the pancreatico-biliary duct, and in defining the morphology of biliary duct cystic dilatations. Ono et al. [48] used cholangiography to define four different abnormal anatomical types of pancreatico-biliary duct junction (PBDJ) and its relevance to the pathogenesis of CCs. Type I APBDJ common union of the pancreatic and common bile ducts occur within the duodenal wall but with separate openings on the duodenal wall; type II APBDJ separate entry of the pancreatic and bile ducts on the duodenal wall; type III common union of the pancreatic and common bile ducts is outside of the duodenal wall; type IV APBDJ the common union between pancreatic and common bile ducts is formed within the duodenal wall with a single opening. From this definition, it is easy to ascribe the pathogenesis of the common channel theory to type III APBDJ. A common channel length of 0.2-1.0 cm or more outside the duodenal wall is significant to allow reflux of pancreatic juice into the common bile duct [49].

Treatment

Choledochocystoduodenostomy, cholecystojejunostomy, choledochocystojejunostomy, intrahepatic cystojejunostomy, choledochocystogastrostomy, cyst unroofing with hepaticojejunostomy, intra hepatic ductoplasty intra hepatic ductoplasty, used to be the conventional surgical procedures. There is immediate relief of symptoms with either of these technique, but the risk of complications and malignancy in the residual cyst can be high, as high as 2.5-26% [50,51]. Anastomotic site

stricture, formation of bile stones, tendency for recurrent cholangitis due to bile stasis are commonly associated internal drainage procedures [52]. Nevertheless, internal or external drainage is still a useful palliative emergency procedure in unstable patients with cholangitis, perforation of the cyst or malnutrition pending a definitive procedure (cyst excision) [53,54].

The aim of surgery is to excise the cyst in Toto, and the establishment of optimal free drainage anastomotic route. As to which technique of anastomosis is optimal after excision is debatable though [55] Hepaticoduodenostomy and Roux-en-Y hepaticojejunostomy were the most optimal of the drainage procedures. These two techniques were suitable for the three cases we managed with uneventful outcome. Hepaticoduodenostomy was performed in one of the patient because the cyst was easily mobilised and there was no pericystic oedema that would risk of injury to the duodenum and pancreatic duct during the mobilisation. Either of the anastomotic technique of choice has its drawbacks. For instance hepaticoduodenostomy has less post-operative adhesive intestinal obstruction, low risk of anastomotic ulcers, it is more physiological and ERCP can still be performed after hepaticoduodenostomy. But there is the risk of reflux gastritis, eosphagitis, metaplastic changes. Which are less likely to occur with hepaticojejunostomy [56]. Overall, the paradigm shift is more toward hepaticojejunostomy, because its complications rates are less 7% compared to hepaticoduodenostomy of 42% [57,58].

When reconstructing a hepaticoenterostomy is it pertinent to create a wide stoma at the hepatic end for the anastomosis; this is to allow free for drainage and reduce the risk of anastomotic stricture [59]. An end-to-end anastomosis minimises the risk of blind loop syndrome, reflux, stone formation, and recurrent cholangitis from bile stasis. The difference in diameter between the calibre of the distal common hepatic duct and the proximal bowel should not exceed ratio of 1:2.5 for ideal end-to-end anastomosis [60,61]. End-side anastomosis is a considerable choice when there is significant calibre difference. The anastomosis should not be sited more than 0.5 cm from the edge of the proximal bowel, to reduce the risk of blind loop syndrome, stone formation, cholangitis.

The ideal level of resection of the distal end of the cyst can be challenging in the absence of intra operative endoscopy to determine the safest level of resection [62]. Because, there is the fear of performing an incomplete excision, injury to the pancreatic duct; nevertheless the resection should be as close as possible to the duodenum at the level of abrupt calibre change or rather the cyst can be opened and content drained then dissected from the sub mucosal plane under clear vision and the stump closed with absorbable suture. At times the cyst can be so inflamed hampering dissection. In such situation mucosectomy is a suggested alternative as was performed in the two other cases [63]. This technique avoids injury to vital structures. Sometimes by injection of normal saline into the cyst wall dissection can be made easy [64]. Finally, before establishing choledochoduodenostomy or choledochojejunostomy the hepatic hilum can be flushed of biliary sludge with normal saline as was done in one of the patients; this will reduce risk of forming hepatobiliary stones from biliary sludge. Roux-en-Y hepaticojejunostomy is the ideal choice for types I and IV choledochal cyst, as often time's excision of the whole choledochus is performed. Types II and III biliary duct cyst are not frequently reported. When encountered the choledochal diverticulum together with the gallbladder should be excised and a Roux-en-Y choledochojejunostomy performed to reduce the risk of malignancy. A transduodenal sphincteroplasty or transverse duodenostomy in the second

or third portion of the duodenum appear the ideal treatment required for the excision of the type III (cholechocele). Hepatectomy is usually required in type V (Caroli's disease) especially if the cyst are localised to a particular lobe of the liver, a times total hepatectomy with liver transplantation in diffuse cystic disease. Patient with type V (Caroli's disease) are often difficult to manage because of recurrent bouts of post-operative complications, particularly cholangitis, cirrhosis.

Currently there is an attempt at laparoscopic excision of biliary duct cysts with considerable benefits; among which are short hospital stay, better cosmesis, and less risk of post-operative adhesive intestinal obstruction [65,66].

Outcome

Complications commonly seen after hepaticoenterostomy are: anastomotic leaks, pancreatic duct injury, intestinal obstructions due to adhesion and intussusceptions, [67] others include anastomotic site ulceration, cancers, liver failure, cholangitis, pancreatitis, and biliary stone. One of the patients with hepatomegally and cirrhotic changes in the liver in our report had persistent fever after surgery which we attribute to be due to cholangitis.

During the short period of follow up, the patients' showed remarkable improvement in their wellbeing. A post-operative ultrasound in one of the patient with hepatomegally was commendable. The hepatomegally improved from 18 cm to 13 cm 3-months after surgery.

The significance of long follow up should be underscored, because the time interval between cyst excision and diagnosis of cancer is still not well established. Mean age of diagnosis of cancer after complete excision of the cyst ranged 15-26 years [68,69].

Conclusion

Presence of complication should not preclude excision of choledochal cyst with hepatoenterostomy especially if the complication is not life threatening to the patient. Because the patient would be much better off if the cyst is excised as evident in all the three cases we managed. But where the excision of the cyst is not feasible a simple palliation in form external or internal drainage should be done, pending a definitive surgery.

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