

# Extrahepatal Cholestasis Due to Choledochal Cyst Type Iv Todani Classification in Two Years Old Girl

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**Abstract:** Choledocal cyst (CC) is a rare congenital cystic dilation of bile duct and can be associated with serious complications including malignancy and inflammation of the surrounding anatomy. Approximately 80% of CC is diagnosed in infants and young children in their first decade of life. Complete surgical resection is recommended in patients with choledochal malformations with excellent outcome. Hereby we aimed to describe diagnosis and management aspects of choledochal cyst. A two years old girl complained intermittent abdominal pain in her upper right quadrant abdomen, accompanied with fever, nausea and vomiting and decrease of appetite. Physical examination, we found jaundice on sclera and palpable liver 2 fingers below the costal arch. Abdominal ultrasound revealed cystic mass in right lower lobe, suggestive of hepatic cyst with inflammation of cystic head of pancreas and minimal free fluid in lumen. Abdominal CT-scan without contrast showed a choledocal cyst classified as Todani Type IV. A cyst in the bile duct was found during surgical procedure. In conclusion choledocal cyst as one of the differential diagnoses in children, which is characterized by intermittent right upper abdominal pain, jaundice and fever. Appropriate surgical measures should be undertaken to avoid complications.

**Keywords:** Children, Choledocal Cysts, Extrahepatal Cholestasis

## 1. Introduction

A Choledochal Cyst (CC) or biliary cyst is a congenital or acquired anomaly affecting the biliary tree. CC involves widening of biliary tree which may affect extrahepatic and/or intrahepatic segments. Biliary cysts are defined as cystic dilation of biliary tracts in single or multiple segments of extrahepatic or intrahepatic bile ducts. In 1959 Alonso-LEJ et al., classified CC into 3 types based on the location of bile duct dilation. In 1977 Todani et al. modified the last classification of CC by adding 2 types of CC. The classification of these five categories is frequently used nowadays [1, 2]. Approximately 80% of CC is diagnosed in infants and young children in the first decade of life. Clear regional variation exists for choledochal cysts, with two-thirds of cases reported in Asia particularly in Japan. The incidence of CC ranges from 1 in

100,000 to 1 in 150,000 individuals in Western countries to 1 in 13,000 individuals in Japan. Choledocal cysts are 4 times more common in women. CC type I and IV have a female-to-male ratio of 4:1 or 3:1 [1, 3, 4].

The exact etiology of choledochal cysts is still unknown, which has led to postulate several theories. The most popular theory is Babbitt's theory, which states cysts develop from an anomalous pancreaticobiliary junction (APBJ). APBJ is when the bile duct and pancreatic duct join proximally 1 to 2 cm to the sphincter of Oddi. This theory is supported by the presence of high amylase level, moreover several studies have reported increased level of phospholipase A2 and trypsinogen in bile. Eighty percent to 96% of pediatric CCs are related to the APBJ. Other pathophysiological mechanistic hypotheses for CC include weak bile duct wall, sustained increase in intrabiliary pressure, inadequate

autonomic innervation, dysfunction of sphincter Oddi and distal obstruction of CBD [1, 2, 5, 6]

There are several classifications for choledochal cysts. The modified Todani classification with 5 types of CC is the most widely used. Type I cysts represent 50% to 80% of CC and characterized by cystic dilatation of the common bile duct. Type II cysts represent 2% of CC and involve diverticular dilatation along the extrahepatic duct. Type III cysts also known as choledochoceles, represent 1.4% to 4.5% of CC and involve intraduodenal cystic dilatation of the distal common bile duct. Type IV cysts represent 15% to 35% of CC and are multiple. Type V cysts, also known as Caroli's disease, represent 20% of CC and involve multiple dilation limited to the intrahepatic biliary tree. Caroli syndrome refers to the presence of type V CC as well as congenital liver fibrosis [5, 7, 8].

Most cases of CC are diagnosed during childhood. [9] CC presents in children as a right upper quadrant mass, abdominal pain and jaundice, known as the classic triad for CC and found in only 20% of cases [9, 10]. Eighty-five percent of children present with 2 of these classic clinical features, most of which include abdominal mass and jaundice. Infants under 12 months of age may present with jaundice, acholic stool and vomiting. Other features of choledochal cysts are cholangitis, pancreatitis and biliary peritonitis due to cyst rupture. Associated congenital anomalies include multiple bile ducts, sclerosing cholangitis, congenital liver fibrosis, pancreatic cysts and annular pancreas. In a national study, congenital cardiac anomalies occurred in 31% of pediatric patients with CC and most commonly manifested in infancy. Biliary malignancy is seen in 10% to 30% CC. Malignancy is rarely seen in pediatric CC [2, 10, 11].

The diagnosis of choledochal cyst is usually achieved by using multimodal imaging including ultrasound, CT and MRI, including MRCP. Ultrasonography has good sensitivity (71% to 97%) for identifying choledochal cysts [12-14]. The sensitivity of CT cholangiography is 90% for diagnosing choledochal cysts but only 64% for characterizing pancreatic duct [1, 14]. MRCP is the gold standard for diagnosing choledochal cysts with sensitivity 90% to 100%. There are no specific laboratory studies for choledochal cysts. [2, 15].

The management approach of choledochal cysts depends on the cyst type and the extension of hepatobiliary pathology. As a rule, all cysts should be resected, and bile flow should be restored. Early surgical excision of CCs is recommended. [16-19]. Caroli disease (Type V CCs) management depends on the extension of the disease. If it is localized or unilobular, then segmental hepatic resection is the best option. If the disease is diffuse or bilobar and symptomatic, then an orthotopic liver transplant is recommended [7]. The prognosis for those whom develop the malignancy is poor, with 5-year survival rate about 5% [20]. In general, excision of choledochal cysts has shown excellent results with incidence-free rate 89% and an overall 5-year survival rate above 90%. Rarely in children, type IVA cysts have the highest rates of complications. However, after cyst excision, the risk of biliary malignancy remains high, even after 15 years of treatment. [8, 20, 22].

We report a case of choledochal cyst Todani Type IV and inflammation of pancreatic head cyst with anemia, transaminitis and hyperbilirubinemia which presented in early childhood. The objective of this case report is to describe clinical and examination aspects of choledochal cyst.

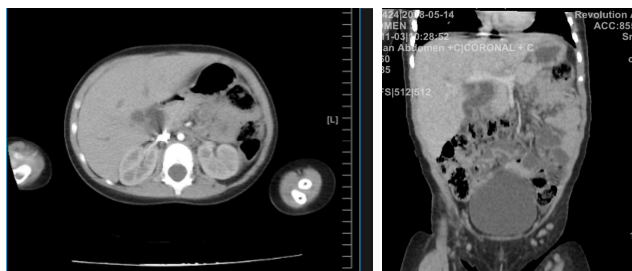
## 2. Case Report

A two-years-old girl was referred from district hospital to emergency unit of Sanglah Hospital with diagnosis abdominal pain, UTI and liver cysts. The patient had chief complaint intermittent abdominal pain since 5 days before admission to the hospital. Pain is felt in the upper right quadrant. The patient also had fever two days ago with highest temperature 38°C. The fever rose up and down with antipyretic medication. She also complained nausea and vomiting with frequency 2 times, in the form of eaten food with volume about 200 ml and poor appetite. History of cloudy urine about 5 days before admission to the hospital. Pain during urinate was denied. History of intermittent urination was denied. Past medical history was denied. History of allergies, surgery and previous transfusions were denied. The family medical history was unremarkable. History of previous hospital treatment were NaCl 0.9% 15 dpm, Ceftriaxone 2x300mg, Paracetamol and Ketorolac. Patient was the second child in the family. No history of congenital anomalies in her family. There was no consanguineous of her parents. No history of illness nor consuming medicine during pregnancy period were noted. There was no abnormality during pregnancy or delivery. Patient had normal growth and development with complete immunization history, before she was sick.

Patient was fully alert when admitted in Sanglah Hospital. The patient's vital signs were pulse 100x/minute, temperature 37.3°C, respiratory rate 22x/minute, oxygen saturation 97% with room air and pain scale 2. The head was normal in shape. There were no sunken eyes, ikteric on sclera, neither conjunctiva injection nor anemia. The pupils light reflex was normal and size were equal. The ears, nose and throat examination were in normal limit. There were no lymph nodes enlargement on the neck or supraclavicular. The chest was symmetrical both on rest and movement. Breath sound was vesicular without rales or wheezing. The first and second heart sounds were normal, regular and no murmur in auscultation. There were no lymph nodes enlargements on both of axilla. Abdomen was not distended and no tenderness. Bowel sound within normal limits. The liver was palpable 2 fingers below the costal arch. Spleen were not palpable. Skin turgor was normal. There were no edema on extremities or lymph nodes enlargement on both of inguinal. Anal and genital examinations were normal. Anthropometric status of patient was 96% (normal) based on Waterlow.

Laboratory tests revealed slight anemia (hemoglobin 10.65), neutrophilia (85.7%), elevated liver enzymes (SGOT 12.8x and SGPT 7x normal value), elevated gamma-GT 11x normal value, elevated bilirubin (total bilirubin 4.8; direct bilirubin 4.72; indirect bilirubin 0.07). Urinalysis revealed yellow color, pH 5, leukocyte esterase +1, nitrite negative,

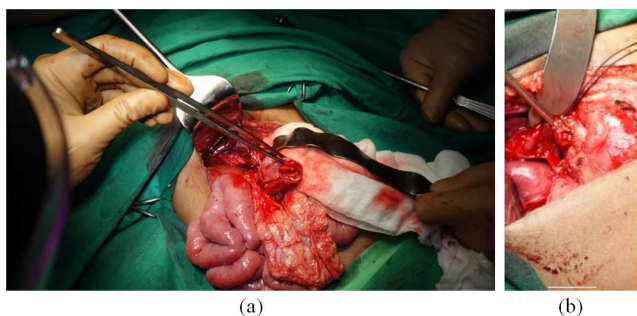
albumin +1, ketone +4, urobilinogen +2, bilirubin +2, erythrocytes 1-2 per field of view, leukocytes 5-8 per field of view, 1-3 flattened cells, with positive bacteria, indicating urinary tract infection. Abdominal ultrasound revealed cystic mass in right lower lobe suggestive of hepatic cyst with inflammation with DD/cystic head of pancreas and minimal free fluid per lumen. Abdominal CT-scan with and without contrast showed a choledochal cyst classified as Todani Type IV (Figure 1) and mild dilatation of the pancreatic duct.



**Figure 1.** Abdominal CT-scan.

Based on the clinical and adjunctive examination, the patient was diagnosed with abdominal pain due to choledochal cyst with inflammation of DD/pancreatic head, transaminitis, cyst with mild anemia normochromic normociter due to chronic infection, well nourished. The patient was given maintenance fluid, ursodeoxycholic acid, paracetamol, parenteral nutrition during fasting and consulted to Pediatric Surgery. Patient was planned to have laparotomy for choledochal cyst excision, hepaticojejunostomy roux en y.

During the surgical procedure, the surgeon found a cyst in the bile duct (figure 2a), then they performed common bile duct cyst excision and hepatic jejunostomy roux en y.



**Figure 2.** (a, b). Cysts in the bile duct and intestinal anastomosis.



**Figure 3.** Bile duct after hepatic jejunostomy roux en y.

Patient was admitted to Pediatric Intensive Care Unit (PICU) after surgery and was given intravenous antibiotics (ceftriaxone and metronidazole). Patient was fasting and supported by total parenteral nutrition for three days. Three days after surgery, tropic feeding from the feeding tube was given. Patient was moved to regular ward and was fully fed on the day ninth of surgery. There were no complications after surgery. The patient was discharged in good condition and being followed up regularly in out-patient clinic.

### 3. Discussion

Choledochal cyst (CC) is a rare congenital cystic dilation of the bile duct and can be associated with serious complications including malignant transformation, cholangitis, pancreatitis and cholelithiasis. CC involves widening of the biliary tree which may affect extrahepatic and/or intrahepatic segments. Approximately 80% of CC is diagnosed in infants and young children in the first decade of life. Clear regional variation exists for choledochal cysts, with two-thirds of cases reported in Asia occurring in Japan. Choledochal cysts are 4 times more common in women. CCs type I and IV have a female-to-male ratio of 4:1 or 3:1 [1, 3, 4]. In this case, patient was two years old girl who had complaints of abdominal pain, UTI and liver cysts without history of congenital anomalies in her family.

The exact etiology of choledochal cysts is still unknown, which led to postulate several theories. The most popular theory is Babbitt's theory, which states that cysts develop from anomalous pancreaticobiliary junction (APBJ). APBJ is when the bile duct and pancreatic duct join proximally 1 to 2 cm to the sphincter of Oddi. This cause increase of pressure, which lead to dilation, inflammation, epithelial damage, dysplasia, and malignancy of the biliary tree. Other pathophysiological mechanistic hypotheses for CC including weak bile duct wall, sustained increase in intrabiliary pressure, inadequate autonomic innervation, sphincter of Oddi dysfunction, and distal obstruction of CBD [1, 2, 5, 6]. In this case, no history of illness nor consuming any medicine during pregnancy period were noted. There was no abnormality during pregnancy or delivery.

CC present in children as right upper quadrant mass, abdominal pain and jaundice also known as the classic triad for CC and found in only 20% of cases [9, 10]. Eighty-five percent of children present with 2 of these classic clinical features, most of which include abdominal mass and jaundice. Infants under 12 months of age may present with jaundice, acholic stool and vomiting. Other features of choledochal cysts are cholangitis, pancreatitis and biliary peritonitis due to cyst rupture. Associated congenital anomalies include multiple bile ducts, sclerosing cholangitis, congenital liver fibrosis, pancreatic cysts, annular pancreas and cardiac anomalies. Biliary malignancy can be found in 10% to 30% CC. Malignancy is rarely seen in pediatric CC [2, 10, 11]. In this case, patient complained intermittent abdominal pain in the upper right quadrant. She suffered nausea, vomiting, decrease in appetite. On physical examination, jaundice on

sclera was found. Liver was palpable 2 fingers below the costal arch was found during physical examination of abdomen. Complaints and signs indicate an association with liver disease.

The diagnosis of choledochal cyst is usually achieved by using multimodality imaging such as ultrasound, CT, MRI, including MRCP. Ultrasonography (US) is the most frequently used imaging modality with low cost, good accessibility and has been shown to be reliable and cost-effective as the sole imaging modality in pediatric population. It has good sensitivity (71% to 97%) for identifying choledochal cysts. US showed a choledochal cyst as a characteristic cystic or fusiform dilatation of the common hepatic duct or intrahepatic duct or occasionally a cyst in the porta hepatis, separate from the gallbladder. It can also indicate associated complications such as cholelithiasis, cholangitis, and malignancy [12-14]. CT cholangiography can identify dilated intrahepatic ducts, distal common bile ducts and pancreatic ducts more accurate than ultrasound, making this modality preferable for type IV and V cysts. The sensitivity of CT cholangiography is 90% for diagnosing choledochal cysts but only 64% for characterizing pancreatic duct [1, 14]. MRCP is the gold standard for diagnosing choledochal cysts with sensitivity 90% to 100%. MRCP is noninvasive, does not expose patient to ionizing radiation nor associated complications such as bleeding, perforation, cholangitis or acute pancreatitis as seen on ERCP. There are no specific laboratory studies for choledochal cysts however the presence of high amylase level and several studies have reported increased level of phospholipase A2 and trypsinogen in bile CCs [2, 5, 15]. Ultrasonography is the only necessary diagnostic imaging. Performing the bile enteric anastomosis in the lower portion of the common hepatic duct is safer and has lower risk of complications [16].

In this case, laboratory tests revealed slight anemia, neutrophilia, elevated liver enzymes, elevated gamma-GT, elevated bilirubin that associated with liver disease and bile obstruction. Patient's abdominal ultrasound revealed cystic mass in right lower lobe suggestive of hepatic cyst with inflammation of cystic head of pancreas and minimal free fluid per lumen. Abdominal CT-scan without contrast showed a choledochal cyst classified as Todani Type IV and mild dilatation of pancreatic duct. Type IV cysts represent 15% to 35% of CCs and are multiple. Furthermore its divided into 2 subgroups. Type IVA involves some dilation affecting both the intrahepatic and extrahepatic biliary tree. Type IVB involves some limited dilation of the extrahepatic biliary tree [1].

The management approach of choledochal cysts depends on the cyst type and the extension of hepatobiliary pathology. As a rule, all cysts should be resected and bile flow should be restored. Type I and type IV CCs are managed with complete excision of the choledochus and restoration of bile flow by preferably Roux-en-Y hepaticojejunostomy (HJ) or hepaticoduodenostomy (HD) [18, 19]. The risk of type II and type III cysts to transform into malignancies is lower. The management of type II consists of simple excision of the cyst or diverticulectomy along with occlusion of the diverticular neck. Type III choledochal cysts (choledochoceles), can be

treated in symptomatic cases or young patients without symptoms by sphincterotomy alone in most cases accompanied by biopsy [7]. Caroli disease (Type V CCs) management depends on the extension of the disease. If it is localized or unilobular, then segmental hepatic resection is the best option. If the disease is diffuse or bilobar and symptomatic, then an orthotopic liver transplant is recommended [20]. In adults and rarely in children, type IVA cysts have the highest rates of complications, such as stones in the intrahepatic tract and stricture formation at the anastomotic site. However, after cyst excision, the risk of biliary malignancy remains high, even after 15 years of treatment. Consequently, it is strongly recommended to follow up long-term with biochemical evaluation and abdominal ultrasound [8, 21, 22]. In this case, patient had laparotomy choledochal cyst excision and hepaticojejunostomy roux-en-y. During the surgical procedure, the surgeon found a cyst in the bile duct, then performed adhesiolysis, cholecystectomy, excision of common bile duct (CBD) cyst, bypass roux en y, liver biopsy, and intestinal anastomosis were performed. There were no complications after surgery. The patient was discharged in good condition and being followed up regularly in out-patient clinic. Another study found that the risk of developing malignancy among patients with choledochal malformation was almost 11 percent and the malignancy risk following cystic drainage surgery was four times higher than that after complete cyst excision. Therefore, complete surgical resection is recommended in patients with choledochal malformations [21].

From this case, we conclude that we should consider choledochal cyst as one of the differential diagnoses in children with symptom of abdominal pain. This awareness may reduce complaints and avoid any delay of definitive surgical treatment. Once choledochal cyst is diagnosed, appropriate surgical therapy should be performed to avoid complications. Type IVA cysts have the highest rates of complications with the risk of biliary malignancy remains high even after 15 years of treatment and it is strongly recommended for long-term follow up with biochemical evaluation and abdominal ultrasound.

## 4. Summary

A two years old girl had intermittent abdominal pain in the upper right quadrant since 5 days before admission, This complaint was accompanied with fever two days ago with highest temperature 38°C, nausea, vomiting, decrease of appetite and history of cloudy urine about 5 days before admission. There was no consanguinity of her parents. From physical examination, we found jaundice sclera and palpable liver 2 fingers below the costal arch on abdominal examination. Laboratory test revealed slight anemia, neutrophilia, elevated liver enzymes, elevated gamma-GT, and elevated bilirubin. Urinalysis indicating urinary tract infection. Abdominal ultrasound revealed cystic mass in right lower lobe suggestive of hepatic cyst with inflammation of cystic head of pancreas and minimal free fluid per lumen.

Abdominal CT-scan without contrast showed choledochal cyst classified as Todani Type IV and mild dilatation of the pancreatic duct. A cyst in bile duct was found during surgical procedure and surgeon performed adhesiolysis, cholecystectomy, excision of common bile duct (CBD) cyst, bypass roux en y, liver biopsy and intestinal anastomosis. There were no complications after surgery. The patient was discharged in good condition and being followed up regularly in out-patient clinic.

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