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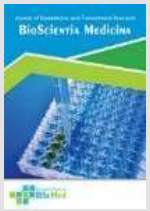

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Multiple Choledochal Cysts: A Case Report

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A B S T R A C T

Background: Choledochal cysts (CC) are congenital dilatations of the biliary tree, characterized by varying degrees of cystic dilatation at various segments of the biliary tract (extrahepatic or intrahepatic), may be detected at any age and in any portion of the bile duct. This study was aimed to present the management of multiple choledochal cyst case. **Case presentation:** A 9 year old boy was brought by his mother to the emergency department at Dr. M. Djamil General Hospital Padang with chief complaint abdominal pain that's getting worse 2 weeks before admission. Abdominal pain first complained by the patient since age of 4, the pain was felt all over stomach area, not localized, not radiated, not corelated with meal time, colicky, relieved with paracetamol. USG results: Choledochal cyst, *Helicobacter pylori* infection, elevated liver enzymes due to suspect suppression of choledochal cyst. Abdominal CT-Scan with contrast results: Intrahepatic and extrahepatic choledochal cyst (type IV A). **Conclusion:** Patients was diagnosed with multiple choledochal cysts, diffuse peritonitis, *Helicobacter pylori* infection, and familial short stature.

1. Introduction

Choledochal cysts are congenital anomalies characterized by cystic dilatation of biliary trees, which can be located at various segments (intrahepatic or extrahepatic). The incidence of this disease is more common in Asian population compared to western population, 1:1000 live births in Japan, and this disease can be detected at any age.[1,2] The pathogenesis of choledochal cysts is still undetermined and still needs further research. Some theories described anomalous pancreaticobiliary union, congenital segmental weakness of the common bile duct wall, or obstruction of the distal common bile duct leading to dilatation. The classic triad of jaundice, abdominal pain and right upper quadrant mass is rare and mostly the clinical presentation is nonspecific abdominal pain, and other symptoms may vary among pediatric population, cholangitis, pancreatitis portal hypertension and liver function abnormalities are also seen.[1,2]

Todani et al., classified choledochal cysts into five types according to the involvement and location of the cysts, and the most common type seen in pediatric is type I (80%-90% of all choledochal cysts which located in extrahepatic biliary tree. The diagnosis of choledochal cysts is made through some imaging studies, initially from ultrasound. Abdominal CT scan can also be performed, and both CT scan and ultrasound are highly sensitive and specific in diagnosing choledochal cysts. The definitive treatment for choledochal cyst is excision surgery depends on the type of the choledochal cysts. Early complications may

include anastomotic leak, wound infection, postoperative leak, and pancreatitis.[1,3,4] This study aimed to present the management of multiple choledochal cyst cases.

2. Case Presentation

A 9 year old boy was brought by his mother to the Emergency Department at Dr. M. Djamil General Hospital Padang with chief complaint abdominal pain that's getting worse 2 weeks before admission. Abdominal pain first complained by the patient since age of 4, the pain was felt all over stomach area, not localized, not radiated, not corelated with meal time, colicky, relieved with paracetamol. Pain seemed to be more frequent in the past two months, interfered patient's activities, not relieved by pain killer. Recurrent vomiting 2 weeks before admission, not related to mealtime, 3-4 times a day, ¼-1/2 cup, contained food, no blood, non-bilious; the last time the patient vomited was two days before admission. Bloated stomach since 1 week ago, patients often felt full, unable to finish meal and felt nausea easily. Constipation since 1 week ago, not bloody but hard consistency, needs laxative for passing stool. Patient looked jaundice noticed by parents 1 week ago along with dark coloured urine. No weight loss. No fever. No history of trying new meals that would cause irritation. No history of trauma of abdominal region. Patient was referred by district hospital with working diagnosis liver cyst based on ultrasound and was given paracetamol to reduce the pain and antibiotics.

Patient is the first out of 3 siblings form his parents' first marriage. Born through spontaneous delivery, birthweight was 3200 grams, birth length 50 cms. The patient received complete immunization until the age of 9 months old, scar BCG (+). The patient began to prone at 4 months of age, sitting at 6 months of age, standing at age 9 months, walking at age 14 months, able to speak at 2 years old, and began to write at 5 years old. The patient lives with parent, brother and sister in a permanent resident. Source of drinking water is from well. Garbage is disposed of by burning in the yard in front of the house. Father's 39 years old, graduated from senior high school, working as labor with salary around Rp.1.500.000,00 per month. The mother is 37 years old and a housewife. The patient had a history of breastfeeding until age of 2 years old, no formula, now having family food 2-3 times a day, with adequate amount of proteins and vegetables, only able to finish half to three quarter of daily portion.

The child was alert and moderately ill. Blood pressure was 90/60 mmHg, heart rate 102 bpm regular, strong pulse, respiratory rate 18 breaths per minute, body temperature 37,2℃, body weight 21 kg, body height 135 cm, weight per age was 56%, height per age was 85% (under percentile 3 CDC curve), height potential ranging from 158-170 cm, upper arm circumference was 13,5 cm, with impression well nourished, and familial short stature. Patient looked jaundice, did not look pale. The skin was warm, no palpable lymph nodes. Head was round and symmetrical, hair was black and strong. Conjunctiva was not anemic, an isochoric pupil with a diameter of 3 mms, light reflex was normal, sclera looked slightly icteric. A nasal flare was not found. Tonsil was not hyperemic and not enlarged, pharynx was not hyperemic. Mucosa of mouth and lips were not dry, not cyanotic. Chest was symmetrical, no retraction, rales nor wheezing. From the heart examination, regular heartbeat, no murmur. There was enlarged abdomen, with abdomen circumference was 45 cm. There was slight abdominal pain locating in epigastric area (Visual Analog Scale 4), the liver was found 3/4-1/2 in size, sharp edge, flat surface, chewy consistency. The spleen was not enlarged. The percussion was tympani, and the bowel sound was normal. There was no shifting dullness. The puberty state was A1G1P1. The peripheral acral was warm, and the capillary refilling time was normal. hemoglobin 11,4 g/dL, white blood cell 6350/mm^3, platelet 443.000/mm^3, differential count 0/5/0/55/33/7, sodium 136, potassium 4,3, chloride 103, PT 10,3 sec, APTT 24,3 sec, total bilirubin 3,3 (3,3x) direct 2,4 (12x), indirect 0,9 (1,5x) Ur 30, Cr 0,7 ALT 184 (4,8x) AST 151 (3,6x).

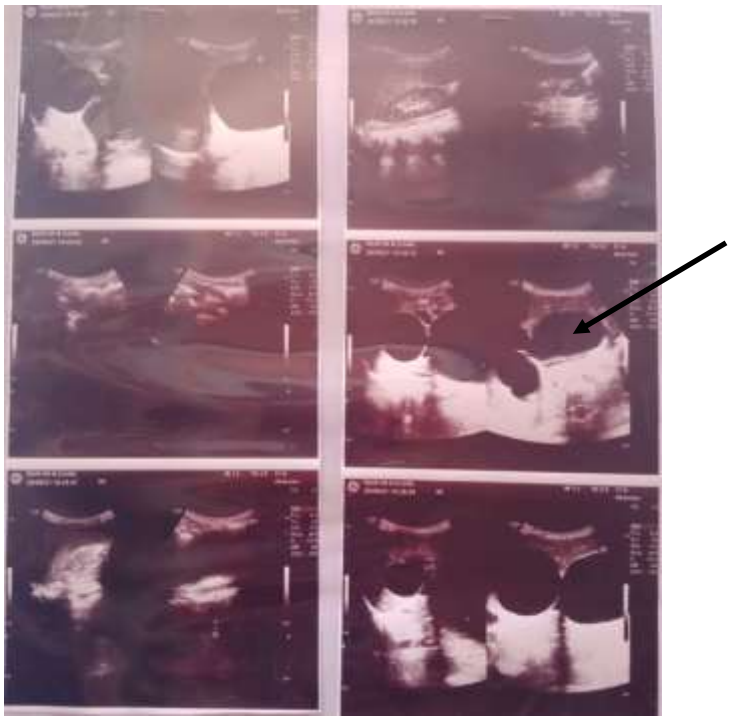

Figure 1. Abdominal ultrasound.

Imaging (Figure 1), Abdominal ultrasound performed on September 28th, 2021: Multiple cyst lesions on liver projection and common bile duct due to suspected choledochal cyst. Impression USG: Choledochal cyst, *Helicobacter pylori* infection, elevated liver enzymes due to suspected suppression of choledochal cyst. Imaging (Figure 2), abdominal CT-scan with contrast results: Intrahepatic and extrahepatic choledochal cyst (type IV A). Impression CT-Scan: Fever et causa suspect mild dehydration, intrahepatic and extrahepatic choledochal cyst, and *Helicobacter pylori* infection.

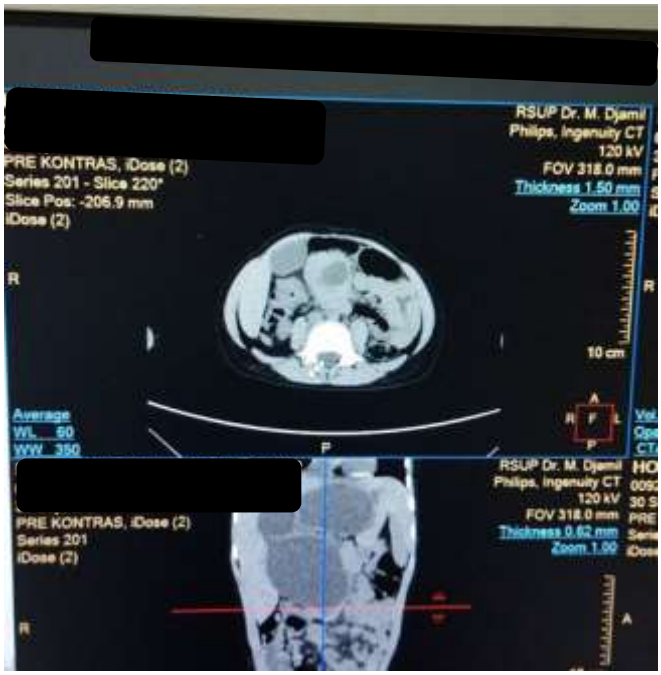

Figure 2: Abdominal CT scan with contrast.

Surgery was performed in general anesthesia, vital signs were stable, and after opening up the operation site, surgeon team found choledochal cyst on cysticus ducts, no cirrhosis and no liver nodul. Drainage of the choledochal cyst was performed, followed by cystectomy and cholecystectomy. The surgery team then decided to perform hepaticojejunostomy end to end and jejuno-jejunostomy end to site leaving drainage on the lateral site of right liver. During the procedure, bleeding was 300 cc, the patient received 200 cc of packed red cells, with fluid balance was +290 cc and diuresis was 3.4 cc/kgBW/hour. The operation was performed for 5 hours, and patient was transferred to recovery room.

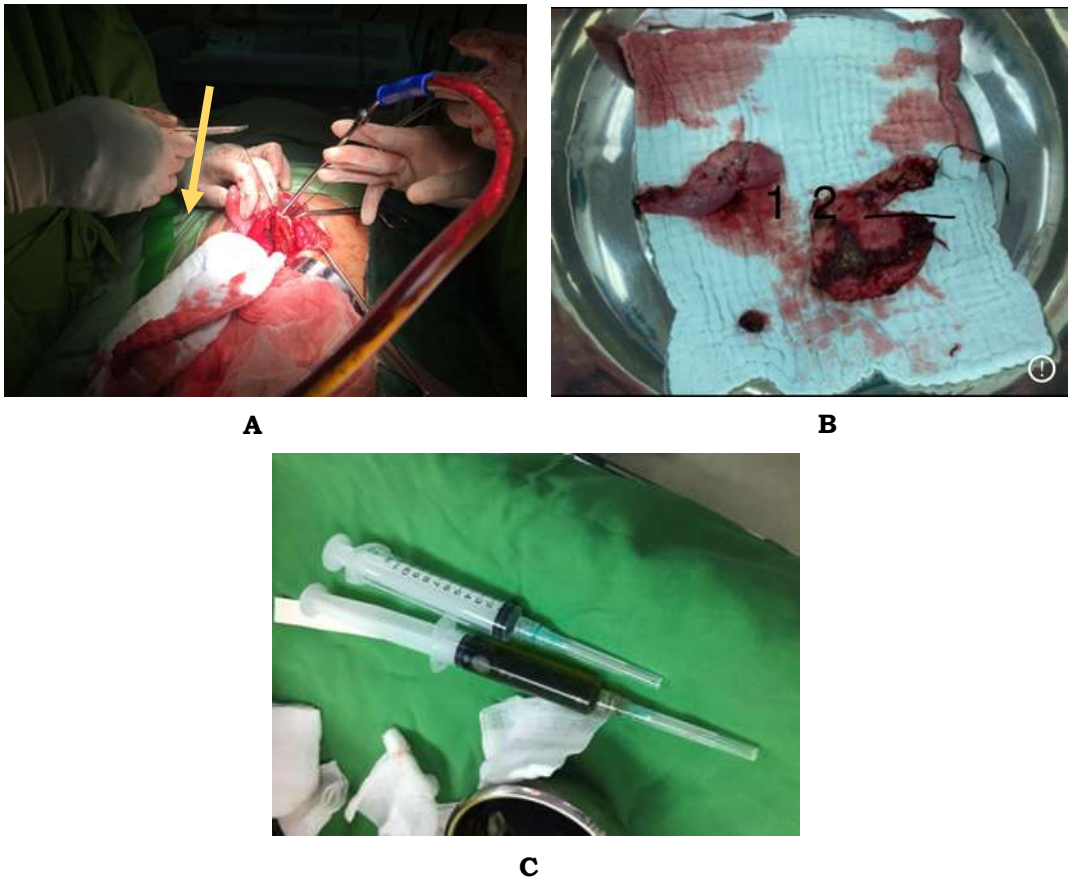

Figure 3. Extrahepatic choledochal cyst; A. Surgical procedure; B. Gall bladder and cyst wall; C. Cyst fluid.

Follow-up post-surgery day 1-3: Post-laparatomy cholecystectomy and drainage of haptic cyst, bypass hepatico-jejunostomy roux en y, *Helicobacter pylori* infection, suspect peritonitis diffuse ec susp leakage of hepato-jejunostomy. Plan: Nothing per oral, total parenteral nutrition, albumin transfusion, correct hypnonatremia within total parenteral nutrition Meropenem 3x800 mg, Metronidazol 2x250 mg iv, Paracetamol 3x250 mg iv omit oral medication temporary, blood and drainage fluid culture, consult to pediatric surgery regarding the bilious drainage production. After consultation, pastient was scheduled for relaparatomy. Relaparatomy day (October 15th, 2021): Post relaparotomy due to bilious leakage, anemia, hypoalbuminemia, and mild hyponatremia. Plan: Intensive care treatment, blood examination after surgery, nothing per oral, total parenteral nutrition, continued intravenous antibiotics, packed red cells, and albumin transfusion. Follow-up day 1-3 post relaparotomy (16th-18th October 2021): Abdominal was not distended, surgery wound was in good condition, no leakage, no bleeding, drainage production was nonbilious and minimal ± 75 cc. Laboratory finding: Hb 10.5 g/dL Leucocyte

13.650/mm^3, IT ratio 0.14 Platelet 384.000/mm^3 Albumin 3.1 g/dL Sodium 138 mmol/L Potassium 4.3 mmol/L Chloride 97 mmol/L Calcium 9.1 mmol/L. Histopathology of the cyst: No malignancy cells were found in this sample. Plan: Continue antibiotics, transfer to the non-intensive ward, increase oral intake, continue treatment for *Helicobacter pylori* infection, follow up for blood culture, urinalysis. Follow up first week after relaparatomy: From abdominal examination the surgery wound was in good condition, abdominal was not distended, bowel sound was normal, and extremity was warm with good capillary refilling time. Some laboratory examinations were carried out to find out the cause of the fever. Laboratory findings: Hb 8.7 g/dL, leucocyte 6.140/mm^3, platelet 462.000 mm^3, IT Ratio 0, MCV 83 MCH 26 MCHC 31, Blood culture: no growth, procalcitonin 0.22 ng/mL serology malaria negative, Tubex negative. Culture of abdominal fluid: Enterococcus faecium, sensitive to Vancomycin. Impressions: Post relaparatomy exploration, abdominal infection due to Enterococcus faecium, normocytic normochromic anemia due to infection. Plan: Definitive antibiotic treatment for the infection. Patient was then discharged 2 weeks after relaparatomy and finishing a course of antibiotics and was advised to have follow up at outpatient clinic.

3. Discussion

A 9 year old boy was admitted to Dr. M. Djamil General Hospital due to abdominal pain and later diagnosed with multiple choledochal cysts, diffuse peritonitis, *Helicobacter pylori* infection, familial short stature. Choledochal cysts (CC) are congenital dilatations of the biliary tree, characterized by varying degrees of cystic dilatation at various segments of the biliary tract (extrahepatic or intrahepatic), may be detected at any age and in any portion of the bile duct. This disease is common in Asian populations, with an incidence of 1 in 13,000 versus 1 in 100,000 in Western populations. There are 4 types of biliary cysts (Type I-V) as proposed by Todani, and this patient was included in type IVA based on abdominal CT scan results. Type IV CC are multiple cysts which can involve both the intrahepatic and extrahepatic biliary tree. Type IV CC can be further subdivided into Type IVa and IVb cysts depending on intrahepatic involvement. Type IVA of CC refers to extrahepatic biliary dilatation with at least one intrahepatic cystic dilatation.[5-8]

The majority (80%) of CC are diagnosed in childhood. Clinical presentation varies and most often consists of nonspecific abdominal pain. The classic triad of jaundice, abdominal pain, and right upper quadrant mass is rare and seen mainly in the pediatric population.[2,9] This patient had been complaining about abdominal pain since the age of four and was suspected of having a cyst at the age of 9 years old. Jaundice was noticed by the mother only once during the admission period and never before. Abdominal quadrant mass was not found in this patient. Tough liver enlargement on physical examination might be misinterpreted. These symptoms were suitable for the manifestation of a choledochal cyst.[10-12]

The diagnosis of CC is typically first accomplished using transcorporeal ultrasound (US). Ultimately, multimodality imaging techniques are often utilized, including computed tomography (CT), magnetic resonance imaging (MRI), and/or endoscopic retrograde cholangiopancreatography (ERCP) to confirm the extent of ductal involvement or the presence of extrahepatic disease. In a retrospective review of 80 pediatric patients who underwent choledochal cyst excision, Jung et al. found that patients with high biliary amylase levels were significantly more likely to be diagnosed later (median age 48 versus 4 months), present with abdominal pain, and had predominantly portal inflammation on histological examination after excision. Abdominal CT scan was carried out in this patient and revealed the multiple choledocal cyst which became a key point diagnosis and in preparing surgery for the patient.[13-15]

The specific approach is largely a function of the type of cyst but generally aims to fully excise the cyst and restore biliary enteric drainage, either primarily into the duodenum or via Roux-en-Y

hepaticojejunostomy (RYHJ). Resection of CC in children is generally well tolerated. In the postoperative period, early complications can include anastomotic leak, postoperative bleeding, wound infection, acute pancreatitis, and pancreatic or biliary fistula. However, most series are without early mortality and report rates of acute complications, including wound infections from 0 to 17%, without significant difference between infants and children. Late complications may include anastomotic stricture, cholangitis, hepatolithiasis, cirrhosis, and malignancy.[6] The use of laparoscopic surgery is more common nowadays compared to open surgery for the management of choledochal cysts. And as of finding the best technique regarding the length of stay, requiring blood transfusion, complication and drainage problem between these two methods are still debatable. A study by Lee et al., which took place in Korean hospitals, showed that the use of a laparoscopic choledochal cyst excision with hepaticojejunostomy is a safe and feasible technique, and the long-term biliary complication was lower compared to open surgery, rendering this a good option for pediatric patients.[3,7,16] This patient underwent laparotomy cholecystectomy along with drainage of the hepatic cyst and bypass hepaticojejunostomy Roux-en-Y.

Several complications that might occur after the excision affect the length of stay and the end results of this disease. Choosing the right technique to minimize the complications is critical. A meta-analysis of 1408 patients (611 in the laparoscopic operation group, 797 in the open operation group) from 7 studies showed that the incidences of complications, including intra-abdominal fluid collection, anastomotic stenosis, bile leak, intrahepatic reflux, cholangitis, pancreatic leak, pancreatic calculi formation, pancreatitis, adhesive intestinal obstruction, Roux loop obstruction, and gastrointestinal bleeding were similar. The laparoscopic cystectomy (LC) group had a significantly lower rate of intraoperative blood transfusion, re-intervention, and adhesive intestinal obstruction. Only one disadvantage of LC is longer operation time. However, it had a shorter duration of recovery of bowel function and hospital stay.[3,7] Some risk factors might affect the outcome of the surgery and also the long term complications such as patient's age at operation, gender, preoperative laboratory findings, presence of preopratove cholangitis and pancreatitis as well as operation time.[16-18]

Spontaneous perforation with biliary peritonitis is a rare complication of bile duct cysts, described in less than 2% of cases, usually in children.[19] The cause of the perforation is thought to be connected with biliary epithelial irritation as a result of reflux of pancreatic juice caused by pancreaticobiliary malunion associated with mural immaturity rather than an abnormal rise in ductal pressure or congenital mural weakness. The diagnosis can be suspected in a patient with acute abdomen, ascites, and dilated biliary tree on imaging.[2,6] This might explain the bilious drainage occured three days after surgery and required relaparatomy in this patient. Patient also complaining about abdominal pain since the age of four, which could also be one of the symptoms of *Helicobacter pylori* infections. Blood samples were sent for examination, and the result was positive for IgG and IgM *H. Pylori*. Patient received first line treatment of *H. pylori* infection which was amoxycillin, metronidazole and omperazole.

Helicobacter pylori is a gram-negative bacterium that selectively colonizes the gastric epithelium and infects over half of the global population.[13,14] Its prevalence varies between countries and among racial groups resident within the same country. It presents with non-specific dyspeptic symptoms with a varying range of severity. In general, children and adults in developing countries are more infected than in developed countries.[20] This may be because of factors such as poor sanitation and lack of potable water supply in developing countries. *H. pylori* infection is causally related to peptic ulcer disease and should be eradicated when detected in such a setting. Possible causal connections have also been made with iron deficiency anaemia (low serum ferritin and hemoglobin levels), short stature, and atopy. *Helicobacter pylori*

infection prevalence was found to be higher in males than females (23.5% vs. 15.4%), though not statistically significant and is similar to findings of other researchers. The highest and lowest prevalence were in age groups 120- 180 months (10-15 years) and 6–59 months, respectively, indicating an increase with age. Several studies reported that the detection of *H. pylori* using serum IgG antibodies displayed 88.4% sensitivity and 93.4% specificity compared with histology.[9,6]

This patient had a concomitant incidence of choledochal cysts and *Helicobacter pylori* infection. It is still unclear which incidence occurred first and whether the first incidence might cause the second incidence to happen. The most reliable explanation regarding to this patient's condition was that the infection of *Helicobacter pylori* might have caused an obstruction of the biliary duct and therefore causing a dilatation which later became the etiology of the choledochal cyst. This condition is supported by the fact that the patient has already complained of abdominal discomfort since the age of 4 years old, and the supporting symptoms regarding choledochal cyst occurred later at the age of 9.[11]

Delayed growth is the most significant nutritional problem around the world, which could lead to long-term effects. The present study suggested that *H. pylori* infection may be a potential risk factor for delayed linear growth in children, even though the specific mechanisms require further investigation. It is still debatable whether delayed childhood growth is due to the direct effects of *H. pylori*-induced inflammation or indirect effects of the infection, e.g. anorexia, abdominal pain, malabsorption or diarrhoea. A study conducted by Yang in 2012 concluded that eradication of *H. pylori* infection promotes growth in children. In conclusion, delayed growth may be due to direct as well as indirect effects of *H. pylori* infection.[9,12,14]

The clinical outcomes of *H. pylori* infection are affected by a number of factors, including virulence, the host gastric mucosa, and the environment. Clinical symptoms of *H. pylori* infection vary between children, with a lower incidence of gastroduodenal ulcers, gastric adenocarcinoma, and mucosa-associated lymphoid tissue lymphoma in children. *H. pylori*-induced gastric inflammation is less severe in children compared with that in adults due to the decreased gastric type 17 T-helper cell/interleukin-17 response in children, which is associated with increased activity of the mucosal regulatory T cells. Therefore, the extra-digestive manifestations of *H. pylori* colonisation in children, including iron deficiency anaemia, cognitive function, type I diabetes mellitus, Henoch-Schonlein purpura, and delayed growth, require constant medical attention. *H. pylori* infection has been reported in numerous studies as a risk factor for delayed childhood growth.[12]

4. Conclusion

A 9 year old boy was brought by his mother to the Emergency Department at Dr. M. Djamil General Hospital Padang with chief complaint abdominal pain that's getting worse 2 weeks before admission. Patients was diagnosed with multiple choledochal cysts, diffuse peritonitis, *Helicobacter pylori* infection, and familial short stature.

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