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Successful Management of Pediatric Todani Type IVa Choledochal Cyst with Roux-en-Y Reconstruction: Diagnostic and Surgical Insights

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A B S T R A C T

Choledochal cysts (CC), or biliary cysts, are congenital dilatations of the biliary tree. Type IVa cysts, involving both intra- and extrahepatic ducts, present diagnostic and management challenges, particularly due to the risk of complications like cholangitis, pancreatitis, and long-term malignancy. Early diagnosis and complete surgical excision with biliary reconstruction are paramount. We report the case of a 4-year-old boy presenting with a six-month history of worsening intermittent upper abdominal pain, nausea, and vomiting. The initial ultrasound suggested a hepatic cyst. Magnetic Resonance Cholangiopancreatography (MRCP) confirmed multiple cystic dilatations involving the intra- and extrahepatic biliary ducts, consistent with a Todani Type IVa choledochal cyst. Laboratory findings showed elevated Gamma GT (179 U/L) but normal bilirubin levels. The patient underwent successful open surgery involving complete excision of the extrahepatic cyst and biliary reconstruction via Roux-en-Y hepaticojejunostomy. His postoperative course was uneventful, with discharge on day six. Follow-up revealed good clinical recovery without immediate complications. In conclusion, Todani Type IVa choledochal cysts are complex anomalies requiring meticulous diagnostic workup and tailored surgical management. This case highlights the utility of MRCP in delineating complex biliary anatomy and reaffirms Roux-en-Y hepaticojejunostomy following complete cyst excision as an effective treatment, offering favourable short-term outcomes in pediatric patients.

1. Introduction

Choledochal cysts (CC), more accurately termed biliary cysts, are rare congenital anomalies characterized by cystic dilatations of the biliary tree. These dilatations can affect single or multiple segments of both the intrahepatic and/or extrahepatic bile ducts. The reported incidence of choledochal cysts varies significantly across geographical regions. Notably, Asian populations, particularly in Japan, exhibit a higher prevalence (1 in 1,000) compared to Western countries, where the incidence is estimated to be between 1 in 100,000 and 1 in 150,000 live births. A significant proportion of cases, approximately 80%, are diagnosed within the first decade of life. The precise etiology of choledochal cysts remains a subject of ongoing investigation. However, the prevailing theory, especially concerning Type I and IV cysts, centers on the presence of an anomalous pancreaticobiliary ductal union (APBDU). This anatomical variation results in the reflux of pancreatic enzymes into the biliary tree. The reflux initiates an inflammatory process, subsequently leading to increased intraductal pressure, weakening of the ductal walls, and the eventual formation of cystic dilatations. The Todani classification system is widely used to categorize choledochal cysts. This

classification is based on the morphology and location of the cysts within the biliary tree. Type I cysts, characterized by fusiform or cystic dilatation of the extrahepatic duct, are the most common, accounting for 50-80% of cases. Type IV cysts, the second most common, are defined by the presence of multiple cystic dilatations.[1-3]

Within Type IV, two subtypes are recognized. Type IVa cysts involve multiple dilatations affecting both the intrahepatic and extrahepatic bile ducts. This is in contrast to Type IVb cysts, which are characterized by multiple dilatations confined to the extrahepatic bile ducts. Due to the involvement of the intrahepatic component, Type IV cysts, particularly Type IVa, are considered more complex in their management. Interestingly, Type IV cysts are reportedly more frequently diagnosed in adults, whereas Type I cysts predominate in the pediatric population. The clinical presentation of choledochal cysts can be highly variable and often non-specific, particularly in older children and adults. The classic triad of symptoms, consisting of abdominal pain, jaundice, and a palpable right upper quadrant mass, is more commonly observed in infants. However, this triad is present in only a minority of older patients. In addition to abdominal pain, which is the most frequent symptom, other symptoms may include nausea, vomiting, and fever. The presence of fever often suggests the development of cholangitis. Symptoms related to complications such as pancreatitis, biliary cirrhosis, portal hypertension, or cyst rupture may also be present. The non-specific nature of these symptoms underscores the necessity for a high index of suspicion and the use of appropriate imaging techniques to establish an accurate diagnosis. Diagnostic imaging plays a crucial role in the evaluation of choledochal cysts. Ultrasound (USG) is frequently employed as the initial investigative modality. It is valuable for detecting ductal dilatation and, in some instances, visualizing the cyst itself. However, Magnetic Resonance Cholangiopancreatography (MRCP) has largely superseded more invasive diagnostic methods. These older methods include Endoscopic Retrograde Cholangiopancreatography (ERCP) and percutaneous transhepatic cholangiography (PTC). MRCP offers several advantages, including excellent visualization of the entire biliary tree, precise delineation of the cyst's extent (including any intrahepatic involvement), and detailed imaging of the pancreaticobiliary junction. Furthermore, MRCP achieves this without the use of ionizing radiation or the risks associated with contrast agents. In contemporary practice, MRCP is considered the gold standard for definitive diagnosis and preoperative anatomical mapping. In this particular case, MRCP was instrumental in confirming the Type IVa classification and guiding the subsequent surgical approach.[4-7]

Choledochal cysts are recognized as premalignant lesions. Patients with choledochal cysts have a significantly elevated lifetime risk of developing cholangiocarcinoma, estimated to be between 10% and 30%, which is substantially higher than that of the general population. This risk increases with age. Additionally, patients with Type I and IV cysts are at a higher risk of malignancy development. Malignancy can arise within the cyst itself or at other locations within the biliary tree. Importantly, malignancy can still develop even years after surgical intervention, particularly if the initial excision of the cyst was incomplete. Given the premalignant nature of choledochal cysts, the standard treatment approach is complete surgical excision of the dilated extrahepatic biliary segments. This includes removal of the gallbladder. Following cyst excision, biliary reconstruction is performed to restore the flow of bile into the intestine. For Type IVa cysts, the management strategy is further complicated by the need to address the intrahepatic component of the disease. Reconstruction of the biliary tree is commonly achieved using a Roux-en-Y hepaticojejunostomy (RYHJ). Although hepaticoduodenostomy (HD) represents an alternative reconstruction technique favored by some centers, RYHJ remains the preferred method, especially in cases with significant inflammation in the hilar region or in cases requiring revision surgery. Timely surgical intervention is

strongly recommended following diagnosis. Early surgery aims to alleviate symptoms, prevent the development of complications, and minimize the long-term risk of malignancy. In infants, early surgical management may also play a role in preventing the progression of liver fibrosis.[8-10] This case report presents a detailed account of a 4-year-old boy diagnosed with a Todani Type IVa choledochal cyst. The report outlines the diagnostic process and the successful surgical management of the condition using open cyst excision and RYHJ reconstruction.

2. Case Presentation

The patient was a 4-year-old male. His origin was confirmed as Madiun, Indonesia. His weight was approximately 16 kilograms, and his height was approximately 100 cms. The patient presented with intermittent upper abdominal pain, nausea, and vomiting as the chief complaints. A detailed history of the present illness revealed that the symptoms had been present for a duration of six months, with a worsening of the symptoms over the last two months. The pain was described as being felt daily in the upper right abdominal region. It is significant to note that the pain was partially relieved by medication, indicating that the patient had likely received some form of symptomatic treatment prior to this presentation. The vomiting occurred approximately two times per day and was characterized as containing digested food. This characteristic of the vomitus suggests that the obstruction or pathology was likely distal to the stomach, allowing for partial digestion to occur before emesis. Associated symptoms were specifically noted to include the absence of a history of fever or jaundice. The lack of fever is an important negative finding, as fever would have suggested an infectious process such as cholangitis, a potential complication of choledochal cysts. Similarly, the absence of jaundice, a yellowing of the skin and sclerae, is a notable negative finding. Jaundice is a common symptom in biliary tract disorders due to the accumulation of bilirubin. However, its absence does not rule out a biliary pathology, as seen in this case. The review of systems included an assessment of the patient's urinary and bowel habits. The patient's urine was described as clear yellow, with normal volume, and without dysuria. Dysuria, or painful urination, was specifically ruled out. Defecation occurred every three days, with soft yellow stool. The stool's consistency and color are relevant, and the frequency of bowel movements provides insight into the patient's gastrointestinal function. The past medical history was significant for the absence of previous similar complaints, a history of jaundice, or prior hospitalizations. This lack of prior medical history is important, as it suggests that the current presentation is likely a new onset of the condition. It also implies that there were no previously identified or treated conditions that could have contributed to the patient's current symptoms. The absence of a prior history of jaundice further reinforces the acute nature of the presentation in terms of biliary obstruction or dysfunction. The general appearance of the patient was described as mildly ill, but the patient was alert and cooperative, with a Glasgow Coma Scale (GCS) of 15, indicated by the term "compos mentis." The assessment of the patient's nutritional status indicated a good nutritional status impression. This is a positive finding, suggesting that the patient's condition, although symptomatic, had not yet significantly impacted their overall nutritional state. The vital signs revealed a heart rate of 80 beats per minute, a respiratory rate of 20 breaths per minute, a SpO_2 of 99% on room air, a temperature of 36.6 degrees Celsius (axillary), and a blood pressure of approximately 95/60 mmHg. These vital signs are generally within the normal range for a 4-year-old child. The heart rate and respiratory rate are within the expected ranges for this age group, and the oxygen saturation is excellent. The temperature is within the normal range, and the blood pressure, although noted as approximate, is also within the physiological parameters for a child of this age. The head and neck examination revealed no scleral icterus and no conjunctival pallor. The absence of scleral icterus, as stated earlier, indicates the absence of jaundice. The absence of conjunctival pallor suggests that the

patient was not anemic. The abdominal examination revealed that the abdomen was soft and non-distended. There was no palpable mass, no hepatosplenomegaly, and no rebound tenderness. The absence of a palpable mass is significant, as choledochal cysts, depending on their size, can sometimes be palpable on abdominal examination. Hepatomegaly, an enlargement of the liver, and splenomegaly, an enlargement of the spleen, were also ruled out. Rebound tenderness, a sign of peritoneal irritation, was also absent. Normal bowel sounds were auscultated. The presence of normal bowel sounds indicates that there was no evidence of an ileus or bowel obstruction. The cardiovascular and respiratory examinations were described as unremarkable, indicating that there were no apparent abnormalities detected in these systems during the physical examination. The laboratory findings included a complete blood count (CBC), liver function tests, and other labs. The complete blood count revealed a hemoglobin level of 13.7 g/dL, a white blood cell count of 12,200/mm³, a platelet count of 427,000/mm³, a hematocrit of 41%, a red blood cell count of 5.08 M/uL, a mean corpuscular volume (MCV) of 80.8 fL, a mean corpuscular hemoglobin (MCH) of 26.9 pg, and a mean corpuscular hemoglobin concentration (MCHC) of 33.4 g/dL. The hemoglobin level is within the normal range for a child of this age, indicating no significant anemia. The white blood cell count is slightly elevated, which could suggest an inflammatory response or infection, although it is not markedly high. The platelet count is within the normal range. The red blood cell indices (MCV, MCH, and MCHC) are within the normal ranges, further supporting the absence of significant anemia. The liver function tests showed a serum glutamic oxaloacetic transaminase (SGOT) or aspartate aminotransferase (AST) level of 31 U/L, a serum glutamic pyruvic transaminase (SGPT) or alanine aminotransferase (ALT) level of 37 U/L, a total bilirubin level of 0.28 mg/dL, and a Gamma GT (GGT) level of 179 U/L. The AST and ALT levels are within the normal range, indicating that there is no significant hepatocellular injury. The total bilirubin level is also within the normal range. However, the Gamma GT level is elevated. GGT is an enzyme primarily found in the liver and biliary tract, and its elevation suggests a potential biliary obstruction or cholestasis, even in the absence of jaundice. This elevated GGT level is a crucial finding in this case, pointing towards a biliary pathology. Other labs included renal function tests, electrolytes, coagulation profile, and C-reactive protein (CRP), all of which were reported as normal. Normal renal function tests indicate that the patient's kidneys are functioning adequately. Normal electrolytes are important for maintaining cellular function and fluid balance. A normal coagulation profile suggests that the patient's blood clotting mechanisms are intact, which is important, especially if surgical intervention is considered. A normal C-reactive protein level suggests that there is no significant acute inflammatory process. However, it's important to note that CRP may not always be elevated in chronic inflammatory conditions. The imaging findings included prior ultrasound results, abdominal ultrasound results upon admission, and magnetic resonance cholangiopancreatography (MRCP) findings. The prior ultrasound, which was performed externally, reported a right hepatic lobe cyst measuring 3.5 x 4 cm. This initial finding raised suspicion for a hepatic cyst, which was a differential diagnosis considered at that time. The abdominal ultrasound performed upon admission confirmed the presence of a cystic lesion related to the right hepatic lobe. The differential diagnosis at this stage included a hepatic cyst versus a choledochal cyst. This highlights the importance of further imaging to differentiate between these two entities. The magnetic resonance cholangiopancreatography (MRCP) was performed, and the findings were crucial in establishing the definitive diagnosis. The MRCP revealed multiple cystic dilatations involving both intrahepatic (left IHBD noted) and extrahepatic (CHD to proximal CBD) bile ducts. The extrahepatic cyst dimensions were approximately 3.3 x 2.6 cm and 3.0 x 2.7 cm. These findings were conclusive for a Todani Classification

Type IVa choledochal cyst. MRCP is the gold standard for diagnosing and classifying choledochal cysts, as it provides detailed anatomical information about the biliary tree and the extent of the cystic dilatations. The identification of multiple cystic dilatations involving both intrahepatic and extrahepatic ducts is characteristic of Todani Type IVa choledochal cysts. The initial diagnosis was a right hepatic lobe cyst. The differential diagnosis included a choledochal cyst. The final diagnosis, based on the comprehensive evaluation and particularly the MRCP findings, was a Choledochal Cyst, Todani Type IVa. This diagnostic progression illustrates the importance of a systematic approach to patient evaluation, starting with initial imaging modalities like ultrasound and progressing to more definitive imaging techniques like MRCP to arrive at an accurate diagnosis. The change in diagnosis from a simple hepatic cyst to a Todani Type IVa choledochal cyst significantly alters the management and prognosis of the patient (Table 1).

The preoperative preparation phase involved several crucial steps to ensure the patient was in optimal condition for surgery. Firstly, a definitive diagnosis of Todani Type IVa Choledochal Cyst was confirmed using Magnetic Resonance Cholangiopancreatography (MRCP). This imaging modality is essential for accurately delineating the anatomy of the biliary tree and the extent of the cystic dilatations, which is critical for surgical planning. Following the diagnostic confirmation, the patient underwent preoperative optimization. This process likely involved a series of standard measures aimed at stabilizing the patient and minimizing potential risks associated with surgery. These measures included ensuring the patient was kept nil per os (NPO), which means nothing by mouth. This is a common practice before general anesthesia to reduce the risk of aspiration. Intravenous (IV) fluid administration was implemented to maintain adequate hydration and electrolyte balance, which are crucial for hemodynamic stability during and after the surgical procedure. Prophylactic antibiotics were also administered. The use of prophylactic antibiotics is a standard practice in surgical cases involving the biliary tract to reduce the risk of postoperative infections, particularly cholangitis. Finally, informed consent was obtained from the patient's parents. Obtaining informed consent is an ethical and legal requirement, ensuring that the parents or guardians understand the proposed surgical procedure, its potential benefits, and its associated risks. The surgical procedure involved a meticulous and multi-staged approach aimed at complete cyst excision and biliary reconstruction. Anesthesia was achieved using general anesthesia, ensuring the patient was completely unconscious and pain-free throughout the operation. The surgical approach was via an open laparotomy. This involves a traditional abdominal incision to access the surgical field. Although minimally invasive techniques are increasingly used, open laparotomy remains a standard approach, particularly for complex cases like Type IVa choledochal cysts. The crucial step was cyst excision. This involved complete excision of the entire extrahepatic choledochal cyst. The dissection was carried out from the hepatic duct confluence, which is the point where the right and left hepatic ducts join, down to the intrapancreatic portion of the common bile duct. This meticulous dissection required careful separation of the cyst from adjacent vital structures, specifically the portal vein and hepatic artery. These are major blood vessels supplying the liver, and injury to these vessels can lead to significant complications. The distal common bile duct (CBD) was ligated close to the pancreas. Ligation involves tying off the duct to prevent bile leakage. Cholecystectomy, the removal of the gallbladder, was also performed. The gallbladder is often removed in choledochal cyst surgery due to its proximity to the cyst and the potential risk of future complications. Reconstruction of the biliary tract was achieved using a Roux-en-Y Hepaticojejunostomy (RYHJ). This is a complex surgical procedure used to restore bile flow from the liver to the small intestine. The Roux limb, a segment of the jejunum (a part of the small intestine), was created. A 40 cm jejunal limb was created, starting approximately 20 cm distal to the

Ligament of Treitz. The Ligament of Treitz is an anatomical landmark used to identify the duodenojejunal flexure. The anastomosis, or surgical connection, between the hepatic duct and the jejunum, was a hepaticojejunostomy. The proximal Roux limb was brought retrocolic, meaning behind the colon, to the porta hepatis, the area where the major vessels and ducts enter the liver. A wide, end-to-side anastomosis was created between the common hepatic duct confluence or hilar plate and the jejunum using fine absorbable sutures. Absorbable sutures are designed to dissolve over time. Another anastomosis, a jejunojejunostomy, was performed approximately 40-50 cm distal to the hepaticojejunostomy. This connects the Roux limb to the remaining jejunum. Regarding the intrahepatic component of the Type IVa cyst, no specific resection was performed. The management strategy focused primarily on extrahepatic cyst excision and the reconstruction of the biliary tract. The decision to not resect the intrahepatic component reflects a common approach in cases where the intrahepatic disease is diffuse or not amenable to localized resection. Drainage was considered, with a surgical drain possibly placed. Surgical drains are sometimes used to remove any fluid collections and monitor for postoperative bleeding or bile leakage. The estimated blood loss during the procedure was minimal, which is a positive indicator of surgical precision and reduced risk of complications. The intraoperative findings documented the observations made during the surgical procedure. There was confirmation of significant cystic dilatation of the extrahepatic bile duct, which aligned with the preoperative imaging findings. Palpable dilatation extending towards the intrahepatic ducts was also noted. This observation is consistent with the Type IVa classification, where both intrahepatic and extrahepatic ducts are involved. The gallbladder was present, as expected, and was removed during the procedure. There were no major intraoperative complications reported, indicating a successful surgical course. Postoperative management focused on supporting the patient's recovery and preventing complications. Initially, the patient was kept nil per os (NPO). This allows the gastrointestinal tract to rest and promotes healing of the anastomoses. The patient received intravenous fluids to maintain hydration and electrolyte balance, and analgesia for pain management. A nasogastric tube was employed briefly for decompression. A nasogastric tube helps to remove gastric secretions and prevent distension. Diet advancement, the gradual introduction of enteral feeding, was initiated as bowel function returned. This is a crucial step in the recovery process, ensuring the patient can tolerate oral intake. Surgical drain management involved removing the drain when the output was minimal. This indicates that the drainage was no longer necessary. Routine vital signs and wound assessment were performed to monitor the patient's overall condition and detect any signs of complications. Liver function tests, including GGT, were monitored, showing a trend towards normalization. This demonstrates the resolution of the biliary obstruction. Routine pain management was provided to ensure the patient's comfort. The immediate outcome was positive, with no early postoperative complications observed. Specifically, there were no signs of anastomotic leak, bleeding, wound infection, pancreatitis, or biliary fistula. An anastomotic leak is a serious complication where the surgical connection between the bile duct and jejunum fails. The patient's condition at discharge was described as good. The patient was discharged on the 6th postoperative day. This relatively short hospital stay indicates a smooth postoperative recovery. The follow-up plan included both short-term and long-term components. The short-term follow-up involved assessment in the outpatient clinic post-discharge. This confirmed continued clinical improvement, complete resolution of presenting symptoms (pain, nausea, vomiting), and satisfactory wound healing. The long-term follow-up is crucial due to the complexity of Type IVa choledochal cysts and the associated risk of malignancy. Lifelong surveillance is recommended. This typically involves regular clinical assessments annually, periodic liver function tests,

and imaging (Ultrasound or MRCP) performed periodically or if the patient becomes symptomatic. The imaging is essential to monitor for potential complications such as anastomotic stricture, intrahepatic stones, cholangitis, or malignancy (Table 2).

Table 1. Summary of patient's clinical findings.

Finding category	Details
Demographics	• Age: 4 years • Gender: Male • Origin: Referred from Madiun, Indonesia • Weight: ~16 kg • Height: ~100 cm
Anamnesis (History)	• Chief Complaints: Intermittent upper abdominal pain, nausea, vomiting. • History of Present Illness: Symptoms present for 6 months, worsening over the last 2 months. Pain felt daily in the upper right abdomen, partially relieved by medication. Vomiting occurred approximately 2x/day, containing digested food. • Associated Symptoms: No history of fever or jaundice. • Review of Systems: Clear yellow urine, normal volume, no dysuria. Defecation every 3 days, soft yellow stool. • Past Medical History: No previous similar complaints, history of jaundice, or prior hospitalizations.
Physical examination	• General: Mildly ill appearance, alert and cooperative (compos mentis). • Nutritional Status: Good nutritional status impression. • Vital Signs: Heart Rate 80/min, Respiratory Rate 20/min, SpO_2 99% (room air), Temperature 36.6°C (axillary). Blood Pressure ~95/60 mmHg. • Head/Neck: No scleral icterus, no conjunctival pallor. • Abdomen: Soft, non-distended. No palpable mass, no hepatosplenomegaly, no rebound tenderness. Normal bowel sounds. • Other Systems: Cardiovascular and Respiratory examinations unremarkable.
Laboratory findings	• Complete Blood Count (CBC): Hb 13.7 g/dL, WBC 12,200/mm^3, Platelets 427,000/mm^3, Hct 41%, RBC 5.08 M/uL, MCV 80.8 fL, MCH 26.9 pg, MCHC 33.4 g/dL. • Liver Function Tests: SGOT (AST) 31 U/L, SGPT (ALT) 37 U/L, Total Bilirubin 0.28 mg/dL. Gamma GT 179 U/L (Elevated). • Other Labs: Renal function tests, electrolytes, coagulation profile, C-Reactive Protein (CRP): normal.
Imaging findings	• Prior Ultrasound (External): Reported as a right hepatic lobe cyst, size 3.5 x 4 cm. • Abdominal Ultrasound (Admission): Confirmed cystic lesion related to the right hepatic lobe; Differential diagnosis included hepatic cyst vs. choledochal cyst. • Magnetic Resonance Cholangiopancreatography (MRCP) (Figure 1): Revealed multiple cystic dilatations involving both intrahepatic (left IHBD noted) and extrahepatic (CHD to proximal CBD) bile ducts. Extrahepatic cyst dimensions approx. 3.3 x 2.6 cm & 3.0 x 2.7 cm. Findings conclusive for Todani Classification Type IVa choledochal cyst.
Diagnosis	• Initial Diagnosis: Right hepatic lobe cyst. • Differential Diagnosis: Choledochal cyst. • Final Diagnosis: Choledochal Cyst, Todani Type IVa.

Table 2. Summary of treatment procedure and follow-up.

Aspect	Details
Preoperative preparation	• Diagnosis Confirmation: Todani Type IVa Choledochal Cyst confirmed via MRCP. • Optimization: Patient underwent preoperative optimization. Standard measures likely included NPO status, IV fluid administration, and prophylactic antibiotics. • Consent: Informed consent obtained from parents.
Surgical procedure	• Anesthesia: General anesthesia. • Approach: Open laparotomy. • Cyst Excision: Complete excision of the entire extrahepatic choledochal cyst, dissected from the hepatic duct confluence down to the intrapancreatic portion of the common bile duct. Careful dissection from adjacent portal vein and hepatic artery. Distal CBD ligated close to the pancreas. • Cholecystectomy: Gallbladder removed. • Reconstruction: Roux-en-Y Hepaticojejunostomy (RYHJ). - Roux Limb: 40 cm jejunal limb created, starting ~20 cm distal to the Ligament of Treitz. - Anastomosis (Hepaticojejunostomy): Proximal Roux limb brought retrocolic to the porta hepatis. Wide, end-to-side anastomosis created between the common hepatic duct confluence/hilar plate and the jejunum using fine absorbable sutures. - Anastomosis (Jejunojejunostomy): Performed ~40-50 cm distal to the hepaticojejunostomy. • Intrahepatic Component: No specific resection performed; management focused on extrahepatic excision and reconstruction. • Drainage: Surgical drain possibly placed • Estimated Blood Loss: Minimal.
Intraoperative findings	• Confirmation of significant cystic dilatation of the extrahepatic bile duct. • Palpable dilatation extending towards the intrahepatic ducts. • Gallbladder present. • No major intraoperative complications reported.
Postoperative management	• Initial Care: Kept nil per os (NPO) initially, received intravenous fluids and analgesia. Nasogastric tube decompression employed briefly. • Diet Advancement: Enteral feeding gradually introduced as bowel function returned. • Drain Management: Surgical drain removed when output was minimal. • Monitoring: Routine vital signs, wound assessment. Liver function tests (including GGT) monitored, showing trend towards normalization. • Analgesia: Routine pain management provided.
Immediate outcome	• Complications: No early postoperative complications observed (anastomotic leak, bleeding, wound infection, pancreatitis, biliary fistula). • Condition at Discharge: Good clinical condition.
Discharge	• Timing: Discharged on the 6th postoperative day.
Follow-up plan	• Short-Term: Assessed in the outpatient clinic post-discharge. Confirmed continued clinical improvement, complete resolution of presenting symptoms (pain, nausea, vomiting), and satisfactory wound healing. • Long-Term: Lifelong surveillance recommended due to Type IVa complexity and malignancy risk. Typically involves: - Regular clinical assessments (annually). - Periodic liver function tests. - Imaging (Ultrasound or MRCP) performed periodically or if symptomatic, to monitor for complications like anastomotic stricture, intrahepatic stones, cholangitis, or malignancy.

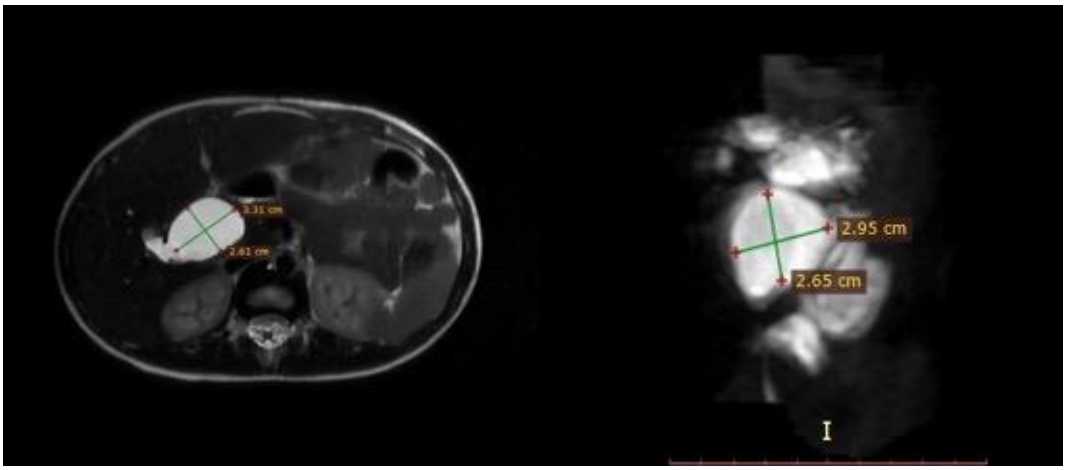

Figure 1. MRCP of the patient.

3. Discussion

Todani Type IVa choledochal cysts, as observed in this case, represent a particularly challenging subset within the spectrum of choledochal malformations. These cysts are distinguished by the involvement of both the intrahepatic and extrahepatic biliary ducts, a characteristic that significantly complicates their management compared to the more common Type I cysts, which are limited to the extrahepatic ducts. The dual involvement necessitates a more intricate surgical approach and a greater emphasis on long-term surveillance. The reported frequency of Type IVa cysts varies, but they constitute a notable proportion of choledochal cyst cases, approximately 15-35%, underscoring the importance of understanding their specific management nuances. The complexity arises not only from the anatomical distribution of the cysts but also from the potential for associated complications and the increased technical demands of the surgical reconstruction, particularly the hepaticojejunostomy. When hilar inflammation or stricturing is present, as can occur in these chronic conditions, the difficulty of creating a patent and functional anastomosis is further amplified. This case underscores the need for a tailored surgical strategy that addresses both the intrahepatic and extrahepatic components of the disease while minimizing the risk of postoperative complications. The management of Type IVa choledochal cysts requires careful consideration of the intrahepatic component, which adds another layer of complexity to the surgical decision-making process. Unlike Type I cysts, where the surgical focus is primarily on the excision of the extrahepatic cyst, Type IVa cysts may present with varying degrees of intrahepatic involvement. The intrahepatic component can range from mild, asymptomatic dilatations to severe, localized saccular dilatations, significant intrahepatic stone burden, biliary strictures, or even suspected malignancy. The optimal management strategy for the intrahepatic component remains a subject of debate and depends on the specific characteristics of the intrahepatic disease in each individual case. In some cases, observation may be a reasonable approach if the intrahepatic dilatations are mild and not causing symptoms. However, in cases with more severe involvement, targeted segmental or lobar hepatic resection may be necessary to address complications or reduce the risk of future problems. In the case presented, the management focused on extrahepatic cyst excision and RYHJ reconstruction, which is a standard approach when widespread hepatic resection is not indicated, suggesting that the intrahepatic dilatation, while present, did not necessitate a major hepatic resection.[11,12]

The patient in this case presented with a six-month history of worsening intermittent upper abdominal

pain, nausea, and vomiting. Notably, the patient did not exhibit jaundice, a classic symptom often associated with biliary tract disorders. This presentation aligns with the understanding that the clinical manifestations of choledochal cysts, particularly in older children and adults, can be non-specific and variable. While the classic triad of abdominal pain, jaundice, and a palpable right upper quadrant mass is more commonly observed in infants, it is present in only a minority of older patients. In this age group, abdominal pain is the most frequent symptom, as seen in this case. Other symptoms may include nausea, vomiting, fever (suggesting cholangitis), or symptoms related to complications such as pancreatitis, biliary cirrhosis, portal hypertension, or cyst rupture. The absence of jaundice in this patient highlights a crucial clinical point, the absence of this sign does not rule out the possibility of a choledochal cyst. The elevated Gamma-glutamyl transferase (GGT) level, despite normal bilirubin levels, served as a critical biochemical clue that pointed towards a biliary tract abnormality. GGT is an enzyme primarily found in the liver and biliary tract, and its elevation suggests potential biliary obstruction or cholestasis, even in the absence of overt jaundice. This case emphasizes the importance of maintaining a high index of suspicion for choledochal cysts in pediatric patients presenting with persistent, non-specific upper abdominal symptoms, even when jaundice is not present. A thorough evaluation, including appropriate laboratory tests and imaging studies, is essential to avoid delays in diagnosis and ensure timely management. The case highlights the challenges in the initial diagnosis of choledochal cysts due to their variable and often non-specific clinical presentation. In this particular case, the initial ultrasound suggested a right hepatic lobe cyst, leading to a differential diagnosis that included a hepatic cyst versus a choledochal cyst. This diagnostic uncertainty underscores the limitations of ultrasound as a standalone diagnostic tool for choledochal cysts, particularly in complex cases. While ultrasound is a valuable initial screening modality for detecting ductal dilatation and sometimes demonstrating the cyst itself, it may not always provide a definitive diagnosis, especially when the anatomy is complex or when the cyst is located in a challenging position. Further, other conditions can mimic the presentation of choledochal cysts, making a high degree of clinical suspicion and the use of more advanced imaging techniques crucial. The differential diagnosis of choledochal cysts can include other cystic lesions of the liver and biliary tree, such as simple hepatic cysts, hydatid cysts, biliary cystadenomas, and Caroli disease. It can also include other causes of abdominal pain, nausea, and vomiting in children, such as gastroenteritis, appendicitis, and intussusception. Therefore, a careful and systematic approach to diagnosis is essential to ensure that choledochal cysts are not missed or misdiagnosed.[13-15]

Diagnostic imaging plays a pivotal role in the evaluation and management of choledochal cysts. While ultrasound is often the initial imaging modality used, it has limitations in fully characterizing the extent of the disease, particularly in cases involving the intrahepatic ducts. In this case, the initial ultrasound suggested a hepatic cyst, and it was the subsequent Magnetic Resonance Cholangiopancreatography (MRCP) that proved to be the key to establishing the correct diagnosis of a Todani Type IVa choledochal cyst. MRCP is now considered the gold standard for the diagnosis and preoperative planning of choledochal cysts. It offers several advantages over other imaging modalities. First, it provides excellent visualization of the entire biliary tree, including both the intrahepatic and extrahepatic ducts. This is crucial for accurately delineating the extent of the cystic dilatations and identifying any associated anomalies, such as an anomalous pancreaticobiliary ductal union (APBDU). Second, MRCP helps in precisely mapping the anatomy of the cyst, including its size, location, and relationship to surrounding structures. This detailed anatomical information is essential for surgical planning as it guides the surgeon in determining the optimal surgical approach and the extent of resection.

Third, MRCP can detect complications associated with choledochal cysts, such as stones, biliary sludge, or even malignancy. Finally, MRCP helps in ruling out other conditions that may mimic choledochal cysts. In this specific case, MRCP was instrumental in confirming the Type IVa classification of the choledochal cyst and in guiding the surgical strategy. The MRCP revealed multiple cystic dilatations involving both the intrahepatic and extrahepatic bile ducts, which is the hallmark of Type IVa cysts. The precise anatomical details provided by the MRCP allowed the surgical team to plan the extent of the cyst excision and the type of biliary reconstruction that would be most appropriate for this patient. The use of MRCP in this case highlights its critical role in the management of choledochal cysts, particularly in complex cases like Type IVa. It has largely replaced more invasive diagnostic methods, such as Endoscopic Retrograde Cholangiopancreatography (ERCP) and percutaneous transhepatic cholangiography (PTC), for definitive diagnosis and preoperative anatomical mapping. These older methods carry a higher risk of complications, such as pancreatitis, cholangitis, and bleeding, and they may not provide as detailed anatomical information as MRCP. MRCP, on the other hand, is a non-invasive imaging modality that does not involve ionizing radiation or the risks associated with contrast agents. It provides excellent visualization of the biliary tree, the cyst's extent (including intrahepatic involvement), and the pancreaticobiliary junction, making it the preferred imaging modality for the diagnosis and management of choledochal cysts in contemporary practice.[16-18]

The cornerstone of management for choledochal cysts, including both Type I and Type IV cysts, is complete surgical excision of the extrahepatic cyst combined with cholecystectomy. This aggressive surgical approach is primarily aimed at mitigating the substantial long-term risk of cholangiocarcinoma, a malignancy of the bile ducts. Choledochal cysts are considered premalignant lesions, with a lifetime risk of cholangiocarcinoma estimated to be significantly higher than in the general population, ranging from 10% to 30%. This risk increases with age and is particularly elevated in patients with Type I and IV cysts. The development of malignancy can occur within the cyst itself or elsewhere in the biliary tree. It is important to emphasize that malignancy can still develop even years after surgical intervention, particularly if the initial excision of the cyst was incomplete. Therefore, the goal of surgery is to remove all of the abnormal biliary tissue that is at risk for malignant transformation. Cholecystectomy, the removal of the gallbladder, is also a standard part of the surgical procedure because the gallbladder is often involved in cystic dilatation and is also at an increased risk of developing malignancy. Incomplete excision or internal drainage procedures alone are associated with significantly higher malignancy rates and ongoing risks of other complications, such as pancreatitis and cholangitis. Internal drainage procedures, which involve creating a communication between the cyst and the intestine without removing the cyst itself, are no longer considered an adequate treatment for choledochal cysts. These procedures leave behind the abnormal biliary tissue, which continues to be at risk for malignant transformation. They also do not address the underlying cause of the cyst formation, which is often an anomalous pancreaticobiliary ductal union (APBDU). APBDU can lead to reflux of pancreatic enzymes into the biliary tree, causing chronic inflammation and further increasing the risk of malignancy. Therefore, complete surgical excision of the cyst is the only way to effectively eliminate the risk of malignancy and other long-term complications.[19,20]

4. Conclusion

In conclusion, this case report illustrates the successful management of a complex case of Todani Type IVa choledochal cyst in a 4-year-old boy. The patient presented with non-specific upper abdominal symptoms, highlighting the diagnostic challenges associated with this condition, particularly in older children where the classic triad of symptoms is often absent. The pivotal role of MRCP in accurately

delineating the complex biliary anatomy and confirming the diagnosis was emphasized, underscoring its superiority over traditional imaging modalities. The surgical management, involving complete excision of the extrahepatic cyst and Roux-en-Y hepaticojejunostomy, proved effective in achieving a favorable short-term outcome. The uneventful postoperative course and the patient's condition at discharge further support the efficacy of this surgical approach. This case also underscores the importance of lifelong surveillance in patients with Todani Type IVa choledochal cysts due to the inherent risk of long-term complications, including malignancy. Regular follow-up with clinical assessments, liver function tests, and imaging studies is crucial for the early detection and management of potential sequelae. This case report contributes to the existing literature by providing further evidence of the successful management of this challenging condition and highlighting key considerations for diagnosis, surgical intervention, and long-term follow-up.

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