Journal of Scientific and Technical Research
11 (1) June 2021, 13-18
http://www.publishingindia.com/jstr

Imaging Diagnosis of a Giant Abdominal Cyst in an Infant

Shabnam Bhandari Grover[1*], Sonali Malhotra[1], Saurabh Pandey[1], Ravi Kale[2] and Anshu Gupta[3]
[1]Department of Radiology, School of Medical Sciences and Research, Sharda Hospital, Sharda University, Greater Noida, Uttar Pradesh, India
[2]Department of Pediatric Surgery, School of Medical Sciences and Research, Sharda Hospital, Sharda University, Greater Noida, Uttar Pradesh, India
[3]Department of Pathology, School of Medical Sciences and Research, Sharda Hospital, Sharda University, Greater Noida, Uttar Pradesh, India
*Corresponding Author: shabnam.grover@sharda.ac.in

Abstract: The usual etiologies of giant abdominal cystic masses in infants are mesenteric cyst, enteric duplication cyst, ovarian cyst in females, cystic lymphangioma, and the presentation of a choledochal cyst in a gigantic form is however, unusual. The primary modality for diagnosis of this entity is ultrasound, followed by MRI. The characteristic ultrasound features of a choledochal cyst are a well-defined cystic lesion which may be replacing any segment of biliary tree and is distinctly separate from the gallbladder. The associated anomalies are biliary atresia, gallbladder atresia, hepatic fibrosis and also anomalies of the pancreatico-biliary system. MRI with MRCP has a conclusive role in confirming the ultrasound diagnosis. Choledochal cysts are classified based on Todani *et al.*'s method as comprising of five types. Herein we report the case study of a 4 month old male infant afflicted with a gigantic Type 1 Choledochal cyst, which was diagnosed by us at the first instance itself by ultrasound examination and was corroborated by MRI. The diagnosis was further confirmed at surgery and histopathology. The recommended treatment of resection of the cyst accompanied by a hepatico-jejunostomy by pass procedure, was successfully performed in the reported infant.

Keywords: Choledochal cyst, Giant abdominal cyst, Infant, MRI, Ultrasound features.

I. Introduction

Abdominal masses in infants can be classified as solid, cystic or those comprising of mixed contents [1]. The etiology of an abdominal mass in an infant, in the order of frequency is likely to be of renal origin in 55% cases, of gastrointestinal tract origin in 15% of cases, of pelvic origin in 15% cases, adrenal masses comprise 10% of cases and a hepato-biliary origin comprises only 5% of all the masses [1]. Amongst the hepatobiliary etiology of cystic abdominal masses in children, hepatic mesenchymal hamartoma, hepato- biliary cystadenoma and choledochal cysts have been reported, but are all rare [2]. Choledochal cyst is a rare congenital malformation of the biliary tract and the gigantic varieties of these are further rare [3-10]. The more frequently reported gigantic abdominal cysts in children have been found to be due to mesenteric cyst, ovarian cyst in females, enteric duplication cyst and cystic lymphangioma both of intra and retroperitoneal location [2].

Ultrasound is the primary imaging modality used to evaluate an abdominal mass in children [11]. Herein we present a case of a gigantic abdominal mass in a 4-month-old male infant which was diagnosed by us using ultrasound evaluation, in the first instance itself as a giant choledochal cyst. The ultrasound diagnosis was corroborated by MR imaging. The imaging diagnosis was found to be accurate and was confirmed at surgery and at histopathology as well.

II. Case Report

A male infant of 4 months was brought by his parents with complaints of increasing abdominal girth, jaundice and worsening respiratory distress for one week. The infant was a child born of non-consanguineous marriage. The birth had occurred at full term, as a vaginal delivery and there was no history of perinatal illness. At current presentation the clinical examination revealed a jaundiced child with respiratory distress and respiratory rate of 40 per minute. The abdomen was distended and there was a large palpable mass in right hypochondrium, which measured approximately 6 x 6 cm in size. The laboratory evaluation revealed Hemoglobin-7.4 g/dl, Hematocrit-24%, Neutrophils-38% and Lymphocytes-

48%. Total bilirubin-7.22 mg/dl, Direct bilirubin-5.01 mg/dl, Indirect bilirubin-2.21 mg/dl, SGOT-171 U/L, SGPT-72 U/L, ALP-1230 U/L. The clinical differential diagnosis was a hepatic tumor versus choledochal cyst or mesenteric cyst and the patient was referred for abdominal sonography for further evaluation. Abdominal sonography was performed on a Philips equipment, model Epiq 7, using a convex probe. Abdominal ultrasound revealed a very large cystic lesion in the sub-hepatic location which measured 8.2 x 8.1 x 7 cm in size. The lesion was in the expected location of CBD and no other structure resembling a CBD was localized. The cyst showed intense posterior acoustic enhancement. The contents of the cyst were few echogenic debris. The cyst was gigantic and resulted in superior displacement of the liver and posterior displacement of the right kidney (Fig. 1). The liver and intra hepatic biliary radicals were however, normal. The gallbladder was localized using a high frequency linear probe and was found to be normal. Minimal fluid was seen in the hepatic sub-capsular region. The pancreas, pancreatic duct and left kidney were normal. The bowel and urinary bladder were normal (Fig. 1). An ultrasound diagnosis of choledochal cyst (Todani Type I) was arrived at. Subsequently an upper abdomen MRI with MRCP examination was performed under sedation, on a Philips MRI (3 Tesla) equipment. The study revealed that CBD was replaced by a large cystic mass located in the subhepatic region, in the expected location of the CBD. The cyst measured 8.2 x 7.2 x 8.1 cm in size. The cyst showed a fluid-fluid level. Antero-laterally the cyst was extending up till the abdominal wall. The cyst was abutting the right kidney and displacing it further posteriorly. Medially the mass was displacing the pancreatic head to the left side. A minimal amount of fluid was seen in the hepatic sub-capsular region. The pancreas, pancreatic duct and left kidney were normal (Fig. 2). The MR appearances corroborated the ultrasound findings and the final imaging diagnosis was Choledochal cyst (Todani Type I). The pediatric surgery team planned for elective surgery after stabilizing the infant with two successive, uneventful blood transfusions. The surgical procedure planned was a cystectomy followed by a Roux-en-Y hepatico-jejunostomy bypass. The incision used was a right sub-costal approach revealed a gigantic cyst in the location delineated by the imaging studies. The cyst was aspirated and allowed to collapse in order to facilitate cystectomy. The aspirated fluid was approximately 350 ml and was dark green in color. The decompressed cyst revealed a tubular structure which measured approximately 9 cm in size and it was resected along with entire extra-hepatic biliary tree. Reconstruction was performed using a Roux-en-Y limb of jejunum with end-to-end anastomosis of the common hepatic duct with the jejunum (Fig. 3). The histopathological examination of the specimen was performed using H & E stain, which confirmed a biliary origin of the cyst (Fig. 4). The histo-pathology examination also revealed hepatic fibrosis (Fig. 4). The post-operative recovery at hospital was managed under intensive care for the first 72 hours and the infant was stable. However, on the fourth day the parents took the patient away against medical advice. Further follow-up was therefore unfortunately not possible.

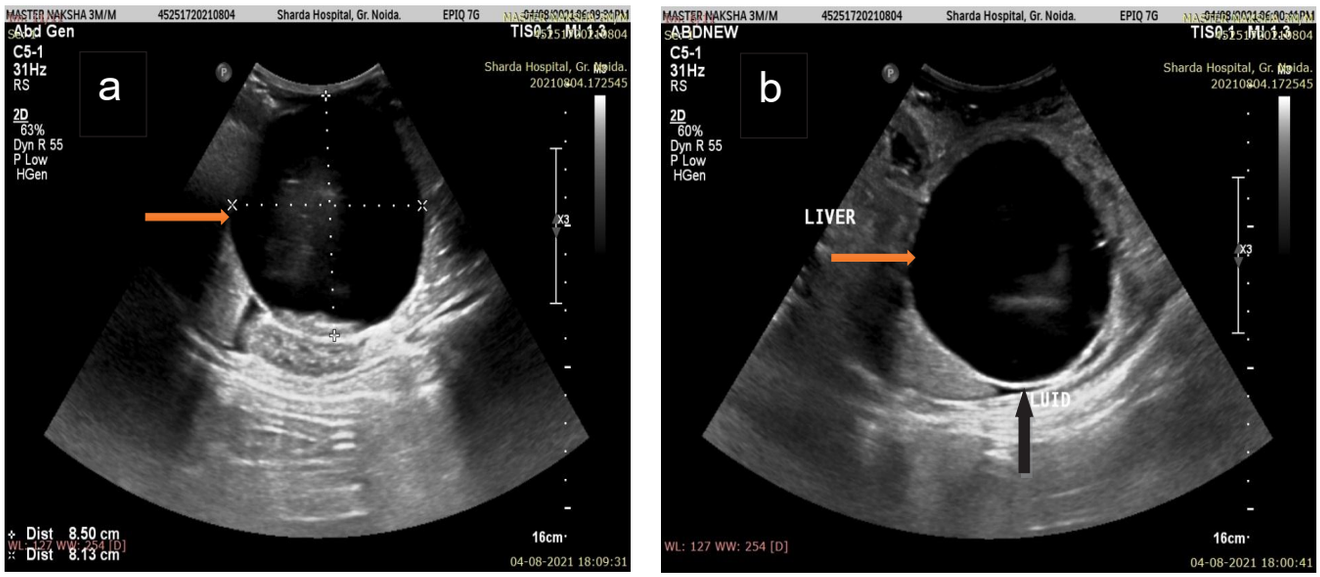

Fig. 1 (a, b): Abdominal ultrasound image in the 4 month male infant shows a gigantic cystic lesion in the sub-hepatic location, which measures 8.2 x 7.2 x 8.1 cm in size (orange arrow, a-b). Minimal fluid was seen in the hepatic sub-capsular region (black arrow, b). The cyst is causing superior displacement of the liver and posterior of the right kidney.

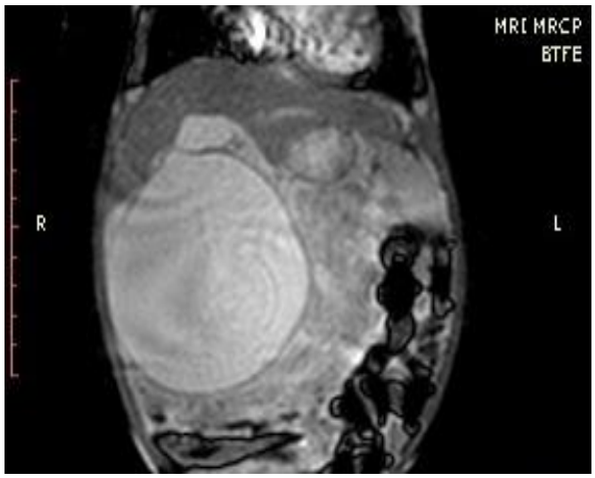

Fig. 2: MRI of upper abdomen in the 4-month infant, BTFE sequence in coronal plane shows a gigantic cystic lesion in the sub-hepatic region. The cyst is gigantic and is indenting the inferior surface of the liver and also causing superior displacement of the organ. Antero-laterally the cyst was seen to be extending up till the abdominal wall; the cyst was abutting the right kidney and displacing it further posteriorly. Medially the cyst is displacing the pancreatic head to the left side.

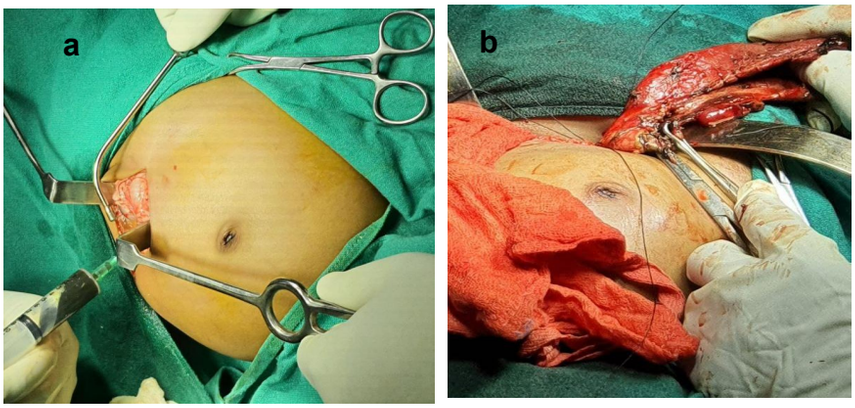

Fig. 3 (a, b): A right sub-costal incision was used for the surgical approach (a). A large cystic lesion was revealed which had to be decompressed before excision (b). The aspirate from the choledochal cyst was a dark green viscous fluid approximately 350 ml, in volume.

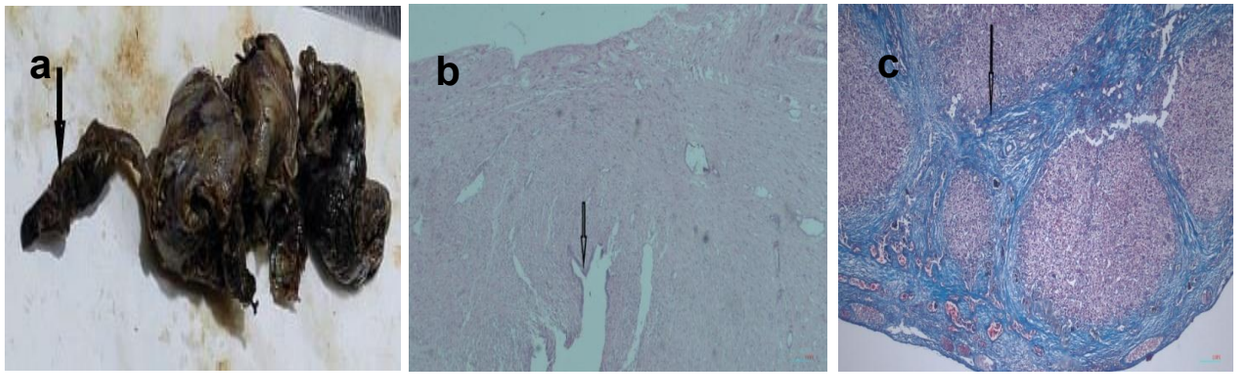

Fig. 4 (a, b, c): Gross specimen of the resected choledochal cyst wall, with attached gallbladder (black arrow, a). Microphotograph of the choledochal cyst which revealed that wall was lined by biliary epithelium (H&E-200x) (black arrow, b). Microphotograph of liver biopsy showed that early fibrosis was present (masson trichrome scanner view) (black arrow, c).

III. Discussion

Choledochal cysts comprise a spectrum of congenital morphological abnormalities of biliary ductal system presenting as cystic dilatation of various segments of this system in different combinations [3-10]. The reported incidence of these anomalies is relatively higher in the Asian population at the rate of 1/1000 births and much rarer in the western population in the rate of 1/100000-1/150000. There is known female preponderance with male: female ratio of 1:3-4 [4, 7]. The earliest reports of this entity are attributed to Vader and Ezlerin 1723 [10]. Alonso-Lez and colleagues are credited with first ever classification of the entity in 1959 [5]. The currently followed classification is attributed to Todani *et al*. which dates back to 1977 [10]. The etiology of the entity has been proposed by various investigators as occurring due to reflux of pancreatic fluid into the biliary tree, due to an anomalous pancreatico-biliary duct union. It is believed that the pancreatic enzymes cause injury and inflammation to the duct walls leading to their dilatation and cyst formation [5, 10]. This theory of reflux is attributed to Babbit who proposed the same vide his publication of 1959 [10].

Choledochal cyst were classified by Todani *et al.* into 5 types (Fig. 5). Type I involves the dilatation of the entire common hepatic or common bile duct or segments of each and it is the most commonly encountered (80-90%) variety of all choledochal cysts. Type I choledochal cysts can be further sub-classified into IA, which is a cystic dilatation of the common bile duct as in the presented case. Type IB is a focal segmental dilatation of the common bile duct and Type IC is represented by a fusiform dilatation of both, the common hepatic and the common bile duct. Diverticular dilatation from the common bile duct occurs in Type II. Type III is a choledochocele, located within the duodenal wall at the pancreatico-biliary junction. Type IV choledochal cysts manifests as multiple cysts which can involve both the intrahepatic and extrahepatic biliary tree. Type IV choledochal cysts can be further subdivided into Type IVA and IVB cysts depending on intrahepatic involvement. Type IVB refers to multiple extra hepatic biliary cysts without intrahepatic involvement. Type V choledochal cyst, or Caroli disease, appears as an intrahepatic cystic dilatation without evidence of extra hepatic dilatation (Fig. 5) [12].

Majority of choledochal cyst are diagnosed in childhood and 80% are known to be diagnosed in the first decade of life [5]. The characteristic presentation of this entity is a unique triad of upper quadrant abdominal pain, palpable mass and jaundice in a young female [5, 10]. Known complications of this entity are pancreatitis, portal hypertension, cholangitis, liver function test abnormalities and obstructive jaundice [5, 8, 10].

A choledochal cyst is typically diagnosed using multi-modality imaging, including ultrasound, computed tomography (CT), and magnetic resonance cholangio-pancreatography (MRCP). Ultrasound is the widely used modality, given its low cost and accessibility, and has been shown to be reliable and cost effective as a single modality imaging in the paediatric population [11]. Characteristic ultrasound features are a well-defined cystic lesion which may be replacing any segment of biliary tree and may show communication with the gallbladder. The cystic lesion shows posterior acoustic enhancement [13, 14]. All known ultrasound features were detected in our patient and the cyst was distinctly separate from the gallbladder. MRI with MRCP has conclusive role in confirming the ultrasound diagnosis. MRI provides a better evaluation of intra hepatic disease and its complications. MRCP is considered as the best imaging modality in pre-operative diagnosis of biliary tree [14]. The case being reported by us qualified to be gigantic Type - I choledochal cyst.

The clinical complications of untreated choledochal malformations range from cholestasis with stone formation to recurrent cholangitis, pancreatitis, biliary and hepatic fibrosis, and malignant transformation (cholangio-carcinoma). It is known that the Type - I choledochal cysts, along with Type - IV cysts, have the highest risk of malignancy [4, 14].

The treatment of the choledochal cyst is surgery and currently the most popular and accepted surgical approach is total excision of the choledochal cyst. It aims to fully excise the cyst and restore biliary enteric drainage into the duodenum via Roux-en-Y hepatico-jejunostomy [6, 8]. The later approach was used in our patient. A laparoscopic approach is also currently being explored as a minimally invasive surgical technique for managing choledochal cysts in children [8].

The prognosis of patients with this anomaly is generally poor as significant insult has already occurred to the hepatobiliary system before the patient presents for medical attention [6]. Delayed treatment may lead to a series of events, such as the liver cell damage, fibrosis of the biliary channels and liver cirrhosis. Therefore choledochal cysts presenting in adulthood have a worse prognosis than those presenting in childhood [8, 11]. In a patient who survives the surgical treatment despite pre-existing hepatic complications, a long-term surveillance for the likely development of malignancy is essential [6].

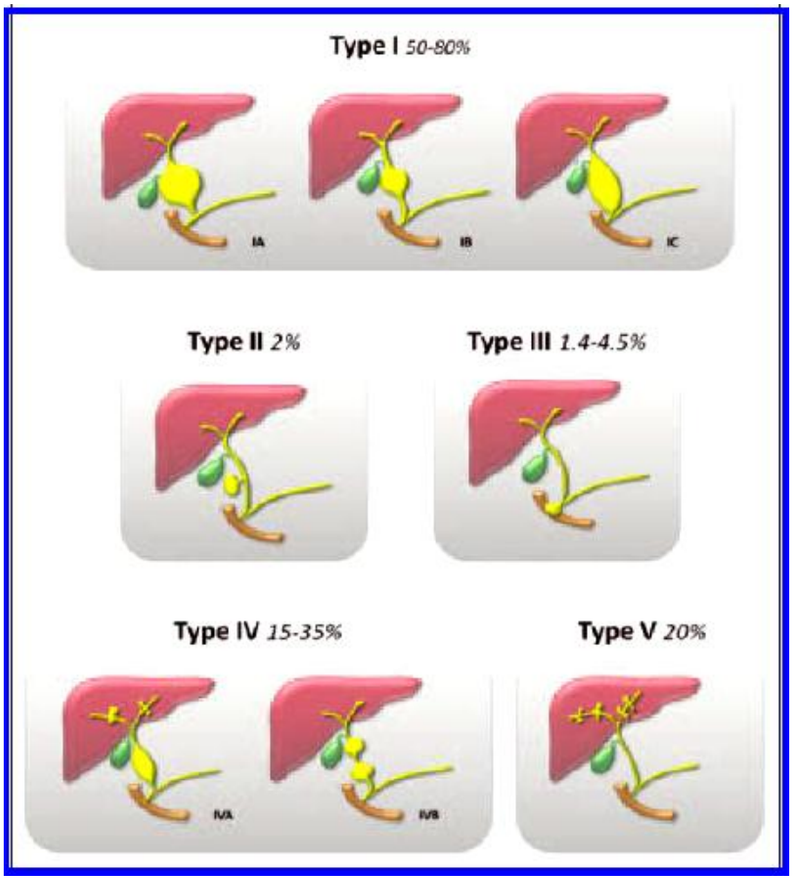

Fig. 5 [12]: The line diagrams depict the Todani modification of the Alonso-Lej classification and relative percentage of occurrence of each type of choledochal cyst. Type - IA, is marked cystic dilatation of the entire extra-hepatic bile duct. Type - IB, is a focal segmental dilatation of the extra-hepatic bile duct, which is usually present distal to cystic duct insertion. Type - IC, comprise a smooth fusiform dilatation of the entire extra-hepatic bile duct. Type - II, is a discrete diverticulum of the extra-hepatic bile duct, while Type - III, is a dilatation of the intra-duodenal segment of the distal common bile duct. Type - IVA, comprises of multiple sites of dilatation of both extra-hepatic and intra-hepatic biliary tree, while Type - IVB, manifests as multiple sites of dilatation only of the extra-hepatic bile duct. Type - V, is recognised by multiple sites of saccular or cystic dilatation of only the intra-hepatic biliary tree (synonym: Caroli disease or communicating cavernous ectasia).

IV. Conclusion

Choledochal cysts are rare anomalies of the biliary tree, which are of unknown aetiology and are typically known to present in young adult females. Presentation in childhood, that too in an early infancy period is very infrequent and its presentation as a gigantic cyst in infancy is rare. The role of radiology and imaging and that of an astute Radiologist is paramount in achieving an early and accurate diagnosis and salvaging this otherwise clinically catastrophic situation. The characteristic imaging features have been highlighted in our report, which we believe will increase awareness of this entity by Radiologists and Clinicians alike.

References

[1] D. E. Light, F. R. Pianki, and E. H. Ey, "Abdominal mass in an infant," *Journal of American Osteopathic College of Radiology*, vol. 4, no. 25, 2015.

[2] L. Ferrero, R. Guanà, G. Carbonaro, M. G. Cortese, L. Lonati, E. Teruzzi, and J. Schleef, "Cystic intra-abdominal masses in children," *Pediatric Reports*, vol. 9, no. 3, p. 7284, 2017, doi: 10.4081/pr.2017.7284. PMID: 29081933; PMCID: PMC5643949.

[3] J. H. Yoon, "Magnetic resonance cholangiopancreatography diagnosis of choledochal cyst involving the

cystic duct: Report of three cases," *The British Journal of Radiology*, vol. 84, pp. e18-e22, 2011.

[4] M. Kumar, and S. Rajagopalan, "Choledochal cyst," *Medical Journal Armed Forces India*, vol. 68, no. 3, pp. 296-298, 2012, doi: 10.1016/j.mjafi.2012.04.011.

[5] K. C. Soares, D. J. Arnaoutakis, I. Kamel, N. Rastegar, R. Anders, S. Maithel et al., "Choledochal cysts: Presentation, clinical differentiation, and management," *J American College of Surgeons*, vol. 219, no. 6, pp. 1167-1180, 2014, doi: 10.1016/j.jamcollsurg.2014.04.023.

[6] M. A. Moslim, H. Takahashi, F. G. Seifarth, R. M. Walsh, and G. Morrisstiff, "Choledochal cyst disease in a western center: A 30-year experience," *Journal of Gastrointestinal Surgery*, vol. 20, no. 8, pp. 1453-1463, 2016, doi: 10.1007/s11605-016-3181-4.

[7] J. S. Banks, G. Saigal, J. M. D'Alonzo, M. D. Bastos, and N. V. Nguyen, "Choledochal malformations: Surgical implications of radiologic findings," *American Journal of Roentgenology*, vol. 210, no. 4, pp. 748-760, 2018, doi: 10.2214/ajr.17.18402.

[8] N. A. Ndoye, B. WelléI, L. Cissé, D. Guèye, C. Diouf, P. A. Mbaye et al., "Choledochalcyst in children in Dakar: Diagnostic and therapeutic aspects," *African Journal of Paediatric Surgery*, vol. 18, pp. 168-170, 2021.

[9] A. K. Dhua, V. Bhatnagar, D. Kandasamy, A. Mitra, A. Ranjan, and A. Varshney, "Giant choledochal cyst in infancy - A rare entity," *Tropical Gastroenterology*, vol. 36, no. 4, pp. 282-283, 2015.

[10] J. M. Suleiman, D. Msuya, R. Philemon, A. Sadiq, P. Amsi, and J. Lodhia, "A giant choledochal cyst: A case reported from Tanzania," *International Journal of Surgery Case Reports*, vol. 81, 2021, Art. no. 105829, doi: 10.1016/j.ijscr.2021.105829.

[11] G. Papaioannou, and J. McHugh, "Investigation of an abdominal mass in childhood," *The British Institute of Radiology*, vol. 16, no. 2, pp. 114-123, 2004, doi:10.1259/imaging/11980186.

[12] S. Ono, S. Fumino, S. Shimadera, and N. Iwai, "Long-term outcomes after hepaticojejunostomy for choledochal cyst: A 10- to 27-year follow-up," *Journal of Pediatric Surgery*, vol. 45, no. 2, pp. 376-378, 2010.

[13] O. H. Kim, H. J. Chung, and B. G. Choi, "Imaging of the choledochal cyst," *Radiographics*, vol. 15, no. 1, pp. 69-88, 1995.

[14] J. S. Banks, G. Saigal, J. M. D'Alonzo, M. D. Bastos, and N. V. Nguyen, "Choledochal malformations: Surgical implications of radiologic findings," *AJR Am J Roentgenol*., vol. 210, no. 4, pp. 748-760, 2018.