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Evaluation of the Kasai Classification in Cystic Biliary Atresia Based on Intraoperative Findings: A Systematic Review of the Literature

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Abstract

Introduction

Biliary Atresia is a congenital condition characterized by bile duct obstruction, leading to liver failure without timely intervention. The Kasai procedure is the standard treatment, but liver transplantation is often required. Current classifications do not fully address the complexity of cystic biliary atresia, a rare variant with unique features. This review proposes a new intraoperative-based classification to improve diagnosis and management.

Methods

A systematic review was conducted in November 2024 across PubMed, ScienceDirect, Scopus, Web of Science, and Google Scholar using the keywords "Cystic Biliary Atresia" OR "Biliary Atresia, Cystic." The search followed PRISMA guidelines, with titles and abstracts screened by two authors and discrepancies resolved by a third. Included studies described preoperative and intraoperative findings of cystic biliary atresia. Articles lacking morphological data or full text were excluded.

Results

The review included 27 studies with 32 cases. Preoperative imaging identified cystic dilation in all cases, with hepatoportoenterostomy performed in 90.6%. Kasai type III predominated (78.1%), with subtype IIIb1 most frequent (53.1%). Cysts were located in the common bile duct (65.6%) or hepatic hilum (28.1%), with an average diameter of 1.9 cm. Findings revealed significant variability in cyst morphology and biliary communication.

Conclusion

Cyst biliary atresia is a rare and complex variant that challenges current classifications. While type III predominates, a more detailed classification is needed to address variability, improve diagnosis, and optimize outcomes.

Keywords: Biliary Atresia; Cystic Biliary Atresia; Biliary Cyst; Choledochal Cyst; Kasai Procedure; Hepaticojejunostomy; Cholestasis

Abbreviations: BA: Biliary Atresia; HPE: Hepatoportoenterostomy; MKC: Modified Kasai Classification; IBA: Isolated Biliary Atresia; SBA: Syndromic Biliary Atresia; CBA: Cystic Biliary Atresia; CMVBA: Cytomegalovirus-associated Biliary Atresia

Introduction

Biliary Atresia (BA) is a progressive congenital disease characterized by sclerosing inflammation and obstruction of the intrahepatic and extrahepatic bile ducts, resulting in biliary flow stasis. Without timely treatment, it leads to progressive liver failure [1]. Its etiology is unknown and heterogeneous, exhibiting significant variability in the degree of destruction of the extrahepatic bile ducts [2]. The standard treatment for BA is hepatoportoenterostomy (HPE), also known as the Kasai procedure. This surgical technique involves the resection of the biliary remnants, the creation of a Roux-en-Y intestinal anastomosis, and the direct connection of the liver to the intestine to restore biliary flow. However, if the procedure fails or if the patient develops cirrhosis, liver transplantation becomes the definitive treatment [3]. Intervention after 60 days of life can accelerate liver fibrosis and significantly worsen outcomes [3,4]. BA is the

most common cause of pediatric liver transplantation worldwide, accounting for approximately 50% of pediatric liver transplants in the United States, [5] 44% in Europe, and 54% in Australia and New Zealand [6,7]. According to the anatomical classification proposed by Kasai [8], BA is divided into four main types. Type I is characterized by partial or complete obstruction of the common bile duct. Type IIa involves obstruction of the common hepatic duct. Type IIb refers to obstruction of the common bile duct, the hepatic duct, and the cystic duct, without affecting the gallbladder. Type III, the most common, is characterized by obstruction of the bile ducts at the hilar level, with no macroscopically visible ducts in the biliary remnants of the hepatic porta. This type poses significant challenges for surgical management [9].

Years later, Kasai introduced a more detailed modification of this classification. The modified Kasai classification (MKC) is organized into three main levels: types, subtypes based on distal bile duct patterns, and subtypes based on hepatic branch patterns at the hepatic hilum. The primary types include Type I, corresponding to common bile duct atresia; Type II, involving hepatic duct atresia; and Type III, affecting the bile ducts at the hepatic hilum. Based on distal bile duct patterns, three categories are identified: patent common bile duct (a_1: patent common bile duct and atretic hepatic duct; a_2: patent common bile duct with aplasia of the hepatic duct); fibrous common bile duct (b_1: fibrous common bile duct with patent or atretic hepatic duct; b_2: fibrous common bile duct with hepatic duct aplasia); and absent common bile duct (c_1: absent common bile duct with patent or atretic hepatic duct; c_2: absent common bile duct with hepatic duct aplasia), in addition to miscellaneous patterns. Finally, the subtypes based on hepatic branch patterns at the hepatic hilum include: α, dilated hepatic branches (internal diameter >1 mm); β, hypoplastic hepatic branches (internal diameter ≤1 mm); γ, absence of bile duct; μ, fibrous hepatic branches; ν, fibrous mass; and o, aplasia of hepatic branches [10].

In 2012, Davenport [11] introduced a clinical classification comprising four BA subtypes: isolated (IBA), syndromic (SBA), cystic (CBA), and cytomegalovirus-associated (CMVBA). Cystic biliary atresia (CBA) is a relatively rare variant, accounting for 5-10% of cases [1]. It is defined as cystic changes in an otherwise obliterated biliary tract, primarily located in the common hepatic duct or the common bile duct [12]. The typical diagnosis of BA generally includes ultrasound, HIDA scintigraphy, and cholangiography. Ultrasound has demonstrated 90% sensitivity in detecting gallbladder abnormalities associated with this condition. The triangular cord sign shows variable sensitivity ranging from 23% to 100%, while sensitivity for identifying cysts in the porta hepatis reaches 25%. [13]. Despite advances in current classifications, particularly the MKC in increasing detail, they fail to adequately capture the complexity of the cystic variant of BA, especially in cases with heterogeneous obliteration of the extrahepatic biliary tracts or functional gallbladders associated with cystic dilations. This study conducts a systematic review of the literature to analyze these morphological variants and proposes a new classification based on intraoperative findings, aiming to improve diagnostic accuracy and guide surgical management in this rare form of the disease.

Materials and Methods

In November 2024, a comprehensive bibliographic search was conducted across the PubMed, ScienceDirect, Scopus, Web of Science, and Google Scholar databases, using the keywords: "Cystic Biliary Atresia" OR "Biliary Atresia, Cystic", with no restrictions on the publication year. Additionally, the references of included studies were manually reviewed to identify any potentially eligible studies. Full-text articles were accessed through institutional accounts with access to restricted resources, including journals and publishers with agreements with our institution. The study followed the PRISMA guidelines [14].

Articles retrieved were initially screened by title and abstract by two authors (F.E.S. and G.R.S.M) to assess their eligibility and inclusion potential. In cases of disagreement, a third investigator (S.T.S.) resolved the discrepancies through discussion and made the final decision. Subsequently, relevant full-text articles were reviewed for the investigation. Studies meeting the following criteria were selected: (1) Publications addressing cystic biliary atresia; (2) Case reports and case series; (3) Cases of patients with complete descriptions of preoperative and intraoperative morphological and anatomical characteristics; and (4) Studies involving human subjects. Articles were excluded if they were published in a language unknown to the authors, lacked an abstract in English, or did not provide access to the full text. Articles were also excluded if they did not include a complete morphological description or, at a minimum, diagnostic or surgical imaging figures that complemented the clinical case description and allowed the authors of this article to classify the cases into at least one subtype of the Kasai classification.

Results

A flow diagram (Figure 1) outlining the study selection process was created. The initial searches in electronic databases yielded a total of 907 studies, and 4 additional studies were manually retrieved from the reference lists of included studies. After removing duplicates, 684 studies remained and were thoroughly screened by title and abstract, with only 104 articles selected for full-text review. Following a full-text evaluation, 77 studies were excluded. We identified 27 studies that met the inclusion criteria for the systematic review, comprising a total of 32 patient cases. The characteristics of the patients from the included studies are presented in (Table 1). Thirty-two cases of patients with biliary atresia associated with extrahepatic cysts were analyzed. Patient ages at diagnosis or surgical intervention ranged from 4 to 126 days, with a mean of 46.2 days and a standard deviation of 28.4 days. The main reported symptoms included persistent jaundice, observed in 100% of cases (32/32), acholia in 87.5%

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(28/32), and hepatomegaly in 12.5% (4/32). Less frequent signs included palpable abdominal masses or acute abdominal distress. Preoperative findings identified cystic dilation at the hepatic hilum in 100% of cases (32/32). Ultrasound scans were widely utilized to visualize cystic lesions, while hepatobiliary scintigraphy (99mTc-IDA) demonstrated absent bile excretion in 75% of cases where it was performed (24/32).

Table 1: Clinical Characteristics of Patients from the Studies Included in the Review.

No.	Author/ year	Age/Sex	Symptoms/ Clinical Presentation	Preoperative Findings	Surgical Technique	Intraoperative Imaging Findings	Kasai Classification	Modified Kasai Classification	Cyst Characteristics
1	[14]	7 days/M	Generalized jaundice	Well-defined cystic lesion in the hepatic hilum communicatin g with the liver.	Hepatojej unostomy with Roux-enY	Cholangiography : Cystically dilated common bile duct with complete atresia of its distal portion and the cystic duct. Hypoplastic but patent bile ducts.	Type I	Type Ib1 β	Cyst located between the common hepatic duct and the common bile duct, in contact with the confluence of a non-patent cystic duct. Cyst proximally communicating.
2	[15]	22 days/F	Acholia	Dilated common bile duct. Subhepatic cyst measuring 29 x 39 x 36 mm	Kasai surgery (Hepatoportoenterostomy)	Cholangiography : Dilated common bile duct due to cystic changes, with no connection to the duodenum	Type I	Type Ib1 β	-Cyst located in the common bile duct - Proximally communicating cyst without connection to the duodenum
3	[16]	26 days/F	Persistent jaundice and acholia	Not reported	Hepatojej unostomy with Roux-enY	Cholangiography : Dilated common bile duct and gallbladder with atresia of the common hepatic duct, narrowing of the intrahepatic biliary system, distal atresia, and no contrast passage to the intestine.	Type III	Type IIIb1	- Cyst located in the common bile duct, in contact with the confluence of patent cystic duct. - Cyst isolated both proximally and distally from bile flow.
4	[17]	39 days/F	Persistent jaundice and acholia	Hepatobiliary scintigraphy revealed no excretion of the isotope in the extrahepatic bile duct or the small intestine	Kasai surgery (Hepatoportoenterostomy)	Cholangiography: Through the atrophic gallbladder, it revealed a common bile duct with complete obstructions	Type III	Type IIIb1	Cyst located in the common bile duct, in contact with the confluence of patent cystic duct. - Cyst isolated both proximally and distally from bile flow.
5	[18]	79 days/F	Persistent jaundice and acholia	Cystic dilation at the porta hepatis associated with other congenital malformations.	Kasai surgery (Hepatoportoenterostomy)	Cholangiography: Communication of the intrahepatic biliary tree with the hepatic duct, complete obliteration of the cystic duct, and absence of the common bile duct with no contrast passage.	Type I*	Type Ic1	Cystic dilation at the porta hepatis in the common hepatic duct. Proximal communication of the cyst with intrahepatic bile ducts.

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6	[19]	18 days/M	Persistent jaundice and acholia	(99mTc-IDA) scan confirmed a large cyst, with no passage of tracer into the duodenum. the cyst communicated with dilated hepatic ducts.	Kasai surgery (Hepatoportoenterostomy)	Intraoperative cholangiogram confirmed the 99mTc-IDA scan findings	Type I	Type Ib1	- Cyst located in the common bile duct, in contact with the confluence of patent cystic duct. - Cyst proximally communicating.
7		45 days/F	Persistent jaundice with hypocholic stools	99mTc-IDA scan showed no passage of tracer into the extrahepatic biliary tract, the cyst, or in the duodenum	Kasai surgery (Hepatoportoenterostomy)	Intraoperative cholangiogram showed a type III-C BA	Type III	Type IIIc*	- Cyst identified at the liver hilum -Cyst isolated both proximally and distally from bile flow.
8		90 days/M	Persistent jaundice and acholia	99mTc-IDA scan showed no passage of tracer into the duodenum.	Kasai surgery (Hepatoportoenterostomy)	Intraoperative cholangiogram showed a type III-C BA	Type III	Type IIIc*	- Cyst identified at the level of the common hepatic duct -Cyst isolated both proximally and distally from bile flow.
9	[20]	11 days/M	Persistent jaundice	A nuclear medicine scan showed no excretion of tracer from the liver.	Kasai surgery (Hepatoportoenterostomy)	Cholangiogram showed a cyst in the porta hepatis that communicated with the gallbladder. There was no flow of contrast in the intrahepatic ducts or into the duodenum.	Type III	Type IIIb1	- The gallbladder communicated directly with the cyst - Cyst located in the common bile duct, in contact with the confluence of patent cystic duct.
10		60 days/F	Persistent jaundice	Abdominal USG showed a 1.3- · 1.2-cm cyst in the porta hepatis	Kasai surgery (Hepatoportoenterostomy)	Cholangiogram done via the gallbladder filled the cyst, but no contrast was seen distally in the duodenum or proximally in the liver	Type III	Type IIIb1	- Gallbladder and cystic duct communicating with a cystic structure in the area of the common bile duct.
11	[21]	70 days/F	Persistent jaundice and acholia	Hepatobiliary scintigraphy using a Tc-99m DISIDA scan showed homogeneous radiotracer accumulation in the liver.	Kasai surgery (Hepatoportoenterostomy)	Cholangiography : The radiographic image revealed a 2.5-cm cystic collection of the contrast material with no connection to the intrahepatic bile duct or intestinal lumen	Type III	Type IIIb1	- Dissection of the cyst revealed a cystic connection to the gallbladder through the cystic duct
12		65 days/M	Persistent jaundice, acholia and hepatomegaly	Hepatobiliary scintigraphy failed to identify the isotope in the gastrointestinal tract by 24 h	Kasai surgery (Hepatoportoenterostomy)	Cholangiography revealed a 3-cm contrast-filled cystic structure with no communication to the liver or the intestine	Type III	Type IIIb1	- The gallbladder communicated directly with the cyst - Cyst located in the common bile duct, in contact with the confluence of patent cystic duct.

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13	[22]	45 days/F	Jaundice; abdominal examination revealed a lump in right hypochondrium	MRCP: cystic lession in extra-hepatic duct with well visalisation of both hepatic duct giving impression of patent duct	Kasai surgery (Hepatoportoenterostomy)	No performance of this imaging test is reported	Type IIb	Type Ib1	- The cyst was located in the subhepatic region and gall bladder was found to be communicating with it - Blind ending cytic lession - Cystic lession in extrahepatic duct with well visalisation of both hepatic duct
14	[23]	31 days/M	Jaundice and hepatomegaly	USG: Cystic lesion (14 × 9 mm) at porta hepatis separate from duodenum without identifiable gall bladder.	Kasai surgery (Hepatoportoenterostomy)	Firm brownish liver, fibrotic porta inclusive of non-visualized right/left hepatic duct, common hepatic duct, and proximal CBD	Type III	Type IIIb1*	- The cyst was located at the site of terminal part of CBD - Blind ending cytic lession
15	[24]	20 days/M	Acholia	Abdominal USG identified a cystic structure measuring 38 mm in diameter in the region of the porta hepatis	Kasai surgery (Hepatoportoenterostomy)	Cholangiogram showing large cystic dilatation and abnormal etiolated, hypoplastic intrahepatic biliary ducts	Type III	Type IIIb1β* (We consider adding 'β' taking into account the presence of hypoplastic intrahepatic biliary ducts)	- Cyst identified at the liver hilum -Cyst isolated both proximally and distally from bile flow.
16	[25]	25 days/F	Jaundice	Ultrasound showed a small gallbladder connected to an extrahepatic cystic structure, which measured 1.4 × 0.6 cm	Kasai surgery (Hepatoportoenterostomy)	Cholangiography : Prompt opacification of the known extrahepatic cyst, which did not show communication with the bile ducts or the duodenum.	Type IIB	Type IIB1	- The gallbladder communicated directly with the cyst - Cyst located in the common bile duct, in contact with the confluence of patent cystic duct.
17		51 days/M	Jaundice	Ultrasound showed a cystic structure anterior to the portal vein measuring 2.3 × 1.6 × 1.8 cm.	Kasai surgery (Hepatoportoenterostomy)	Opacification of a small gallbladder connected to a large cyst. Sustained contrast injection distended the cyst. No communication with the bile ducts or the bowel was demonstrated.	Type III*	Type IIIb1	- Cystic dilatation of the common bile duct without communication to the duodenum and a fibrotic hepatic duct proximally. - The gallbladder communicated directly with the cyst

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18	[26]	90 days/M	Persistent jaundice	Ultrasound performed that showed a cyst located within the hepatic hilum. Along the anterior wall of the right portal vein, the "triangular cord sign."	Kasai surgery (Hepatoportoenterostomy)	Cholangiography: distal segment of the irregular gallbladder and cystic duct communicating with an approximately 2-cm round cyst in the hepatic hilum. There was no communication with the intrahepatic bile ducts or with any additional extrahepatic bile ducts or bowel	Type III	Type IIIb1	- The gallbladder communicated directly with the cyst - Cyst located in the common bile duct, in contact with the confluence of patent cystic duct.
19	[27]	30 days/M	Jaundice	Ultrasound examination, a 26 × 25 mm cyst was observed at the hepatic hilum	Kasai surgery (Hepatoportoenterostomy)	Cholangiography : atrophy of the gallbladder, with no patency of either the intrahepatic duct or common bile duct	Type III*	Type IIIb1	- Cyst identified at the liver hilum -Cyst isolated both proximally and distally from bile flow.
20	[28]	81 days/F	Acute abdominal distress	Abdominal USG: presence of massive ascites and a cystic lesion at the hepatic hilum measuring 3 cm in diameter, with collected fluid and debris	Kasai surgery (Hepatoportoenterostomy)	Cholangiography : cystic dilation of the proximal common bile duct without contrast passage to the loops, dilation of the common hepatic duct with nearly complete atresia of the right and left hepatic ducts.	Type III	Type III a1, μ* (reported by the article, we consider IIId μ given the permeability of the proximal common bile duct and distal hepatic duct)	- Cyst in the proximal common bile duct in contact with the confluence of a cystic duct and a permeable gallbladder. - Dilation extends through the proximal common hepatic duct with no communication beyond that point.
21	[29]	60 days/M	Persistent jaundice, acholia and hepatomegaly	USG: well-defined cystic lesion at porta hepatis measuring 0.9x1.2 cm.	Kasai surgery (Hepatoportoenterostomy)	No performance of this imaging test is reported	Type I	Type Ib	- Cystic biliary atresia with evidence of obliteration of distal common bile duct and dilated proximal biliary system.
22	[30]	22 days/M	Jaundice and acholia	Abdominal USG identified a gall bladder measuring 20 × 10 mm and revealed a cystic structure measuring 10 mm in diameter in the hepatic hilum	Kasai surgery (Hepatoportoenterostomy)	Cholangiography : only cystic dilatation of the common bile duct connected to the gall bladder was imaged, but the intrahepatic bile duct, pancreatic duct, and duodenum were not imaged	Type III	Type III d γ*	- The gallbladder communicated directly with the cyst - Cyst in contact with the confluence of patent cystic duct.

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23	[31]	4 days/M	Jaundice and acholia	Ultrasonography revealed a cystic mass, 43 × 32 mm in size, at the porta hepatis, without dilatation of the intrahepatic bile duct.	Kasai surgery (Hepatoportoenterostomy)	Cholangiography : tree pattern of intrahepatic bile ductules, a cystic common bile duct, and no connection to the intestinal lumen	Type I	Type Ib	- Large cyst occupying the entire course of the common hepatic duct, from the confluence of the right and left hepatic ducts to the confluence of the common bile duct and cystic duct. - The gallbladder communicated directly with the cyst
24	[32]	126 days/F	Jaundice and acholia	Ultrasonography showed distended gall bladder with two cysts at porta and multiple intrahepatic cysts.	Kasai surgery (Hepatoportoenterostomy)	The contrast was injected into one of the cysts that did not show any communication with the other cysts, intrahepatic biliary radicals or duodenum	Type III	Type IIIb1	- One large cyst was seen at the common hepatic duct -Cyst isolated both proximally and distally from bile flow.
25	[33]	18 days/F	Acholia	An abdominal ultrasound demonstrated an extrahepatic cystic structure measuring 2.5 × 1.7 cm that appeared to be blind-ending	Hepaticoduodenostomy	Cholangiogram: patent gallbladder with distal opacification into a cystic, blind ending structure, with no flow into the duodenum. Proximally, contrast showed communication to two separate branching right and left hepatic ducts and further to non-dilated intrahepatic biliary radicles	Type I*	Type Ib1	- A blind ending cyst involving the CBD was identified in the hepatoduodenal ligament, adjacent to a serpiginous appearing gallbladder. - The gallbladder communicated directly with the cyst
26	[34]	60 days/F	Jaundice and acholia	Abdominal ultrasound demonstrated a cystic lesion in the porta hepatis as well as a probable triangular cord sign and an irregular walled distended elongated gallbladder.	Kasai surgery (Hepatoportoenterostomy)	Image shows opacification of the gallbladder, cystic duct and the cystic lesion documented on the ultrasound. There was no passage of contrast media to the intrahepatic duct nor passage to the duodenal lumen.	Type IIb	Type Ib1	- The gallbladder communicated directly with the cyst - Cyst in contact with the confluence of patent cystic duct.
27	[35]	30 days/F	Jaundice, acholia, excessive cry and fever	MRCP: showed approximately 14 mm × 13 mm-sized cystic lesion in hepatic hilar region in the location of common hepatic duct	Kasai surgery (Hepatoportoenterostomy)	No performance of this imaging test is reported	Type III*	Type IIIb1	- Cystic dilatation at the confluence of atretic common hepatic duct, cystic duct, and common bile duct -The cyst did not communicate with the ducts.

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28	[36]	40 days/M	Jaundice and acholia	MRCP demonstrated a saccular, lobular 2.1 cm cystic structure that appeared to originate from the common bile duct	Kasai surgery (Hepatoportoenterostomy)	Cholangiogram: cystic dilation of the common bile duct with no drainage of bile into the small bowel and minimal reflux of bile into the left and right hepatic ducts	Type III	Type IIIb1	- The gallbladder communicated directly with the cyst - Cyst located in the common bile duct, in contact with the confluence of patent cystic duct.
29	[37]	56 days/F	Jaundice and acholia	MRI of the abdomen showing a large cystic mass in the hilum of the liver	Kasai surgery (Hepatoportoenterostomy)	Cholangiogram: no communication with the intrahepatic bile ducts or with any additional extrahepatic bile ducts or the bowel.	Type III	Type IIIb1	- Cyst identified at the level of the common hepatic duct -Cyst isolated both proximally and distally from bile flow.
30	[38]	60 days/F	Persistent jaundice	The ultrasound found an anechoic cystic lesion along the portal triad which did not show doppler signal	Kasai surgery (Hepatoportoenterostomy)	Cholangiogram was performed by injecting contrast into the atretic gallbladder, showing no flow of contrast into the intrahepatic system nor the small bowel	Type III	Type IIIb1	- Cyst showed no communication with the biliary tree or small bowel. - The cyst was located at the CBD
31	[39]	38 days/F	Jaundice and acholia	(MRCP) showed a CHD cyst (0.7 × 0.5 × 0.7) cm with intermittent development of the CBD	Kasai surgery (Hepatoportoenterostomy)	The liver exhibited high fluorescence, with limited fluorescence in the CHD and no fluorescence in the gallbladder and CBD one hour after ICG injection	Type IIa	Type IIb1	- Cyst identified at the level of the common hepatic duct -Cystic biliary atresia with evidence of obliteration of distal common bile duct
32	[40]	91 days/F	Persistent jaundice	Ultrasound image: cystic dilation of the common bile duct; the gallbladder was not identified.	Kasai surgery (Hepatoportoenterostomy)	Cholangiogram: patency of the biliary ductules in the absence of a normally formed biliary tree.	Type I*	Type Ib1	- Cyst showed no communication with the biliary tree or small bowel. - The cyst was located at the CBD

M: male, F: female, MRCP: Magnetic Resonance Cholangiopancreatography, MRI: Magnetic Resonance Imaging, ICG: Indocyanine Green, CBD: Common Bile Duct, CHD: Common Hepatic Duct, USG:

Ultrasonography, 99mTc-IDA: Technetium-99m Iminodiacetic Acid.

*The article explicitly indicated the Kasai classification, whether original or modified.

Regarding surgical procedures, hepatoportoenterostomy (Kasai procedure) was performed in 90.6% of cases (29/32). Additionally, two hepaticojejunostomies (6.3%) and one hepaticoduodenostomy (3.1%) were conducted. Intraoperative cholangiography was performed in 90.6% of cases (29/32), confirming biliary obstruction and characterizing the morphology of the cysts. Not all studies reported Kasai classification; however, all cases were reevaluated using available information to resolve any discrepancies in those that did report it. Based on additional images and descriptions, the closest classification was determined. The distribution of Kasai classification was as follows: type I in 15.6% (5/32), type II in 6.3% (2/32), with type IIa and type IIb representing one case each, and type III in 78.1% (25/32). Among the modified subtypes, subtype IIIb1 was the most common, accounting for 53.1% (17/32). Common cyst characteristics included their location, predominantly in the common bile duct in 65.6% of cases (21/32) and at the hepatic hilum in 28.1% (9/32). The average cyst diameter was 1.9 cm, ranging from 0.7 to 4.3 cm. Regarding communication, 46.9% of the cysts showed a connection to the gallbladder (15/32), while 53.1% were isolated both proximally and distally (17/32).

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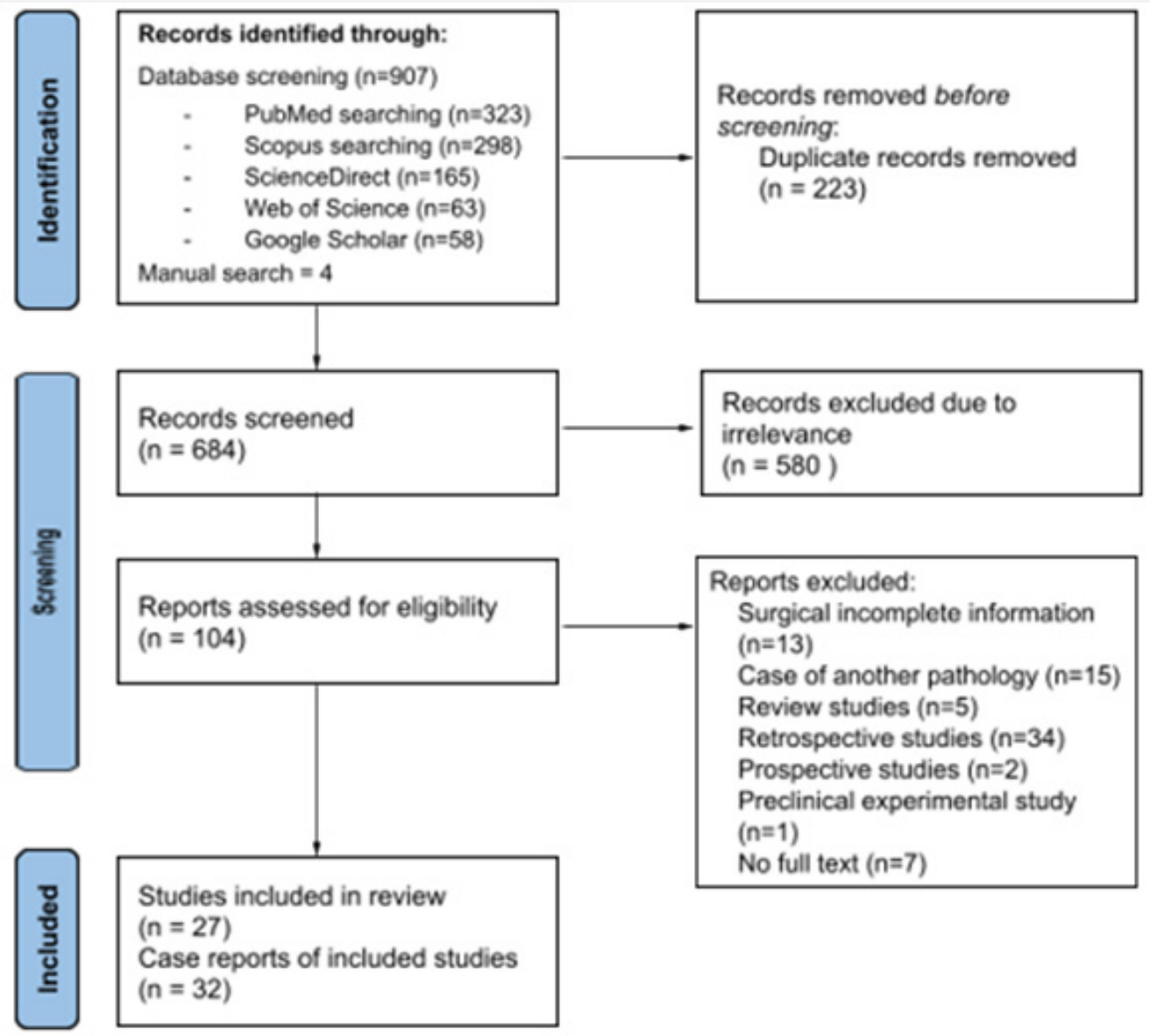

Figure 1: Flow chart of the literature search, identification and selection of the studies.

Discussion

The presence of a cyst at the hepatic hilum, along with cholestasis, presents a critical differential diagnosis with choledochal cysts. Although both entities share some clinical features, they differ in etiology, progression, and treatment. Choledochal cyst dilation, also known as choledochal cyst, is a congenital malformation involving abnormal dilation of the bile ducts. Unlike biliary atresia, choledochal cysts are not exclusively neonatal diseases and are managed variably depending on the cyst type and presentation. Common variants often require resection followed by hepatoenterostomy [41]. Differentiating between these entities using imaging studies and intraoperative findings is crucial to prevent treatment delays, minimize the risk of advanced liver damage, and improve long-term outcomes [2].

The original and modified Kasai classifications were designed to describe conventional scenarios of biliary atresia, characterized by uniform obliteration of the bile ducts. These classifications are primarily based on the level of anatomical and functional disruption of the biliary system [42]. However, significant limitations arise when applying these classifications

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to more complex anatomical variants, such as biliary atresia with cystic dilations-a less common but clinically significant subtype in the differential diagnosis of persistent neonatal jaundice and cholestasis [43]. One of the main challenges is the inflexibility of current categories in addressing the presence of cystic structures. These dilations may be partially or entirely connected to the proximal or distal biliary system or may remain isolated [44]. This variability is inadequately addressed in current classifications, potentially leading to inaccurate or less clinically useful categorizations. Additionally, the Kasai classifications do not explicitly address the functionality of structures like the proximal bile duct, cystic duct, or gallbladder when they are adjacent to or connected with cystic dilations.

Biliary atresia type I accounts for approximately 5% of cases, type II for about 2%, and type III for more than 90% [45]. Our study observed a similar distribution, with type III being the most frequent, followed by type I and type II.

In cases of cystic biliary atresia, evaluating the biliary tree's status is essential to characterize the distribution and drainage toward the dilations. This helps determine whether structures communicate with the bile ducts or are entirely isolated, potentially serving as a prognostic factor for severity. Cysts with proximal communication or patency have a lower likelihood of complete obliteration at the hilum and intrahepatic ducts, reducing the risk of hepatoportoenterostomy failure and the need for transplantation [46]. The Modified Kasai Classification subtypes present limitations in this regard, as they do not account for common anatomical combinations in these cases. Furthermore, they tend to detail less clinically relevant factors, such as fibrosis (μ), hypoplasia (β), narrowing (o), or agenesis of intrahepatic ducts (γ), which are challenging to characterize surgically. At the same time, they superficially address more prevalent anatomical features, such as cysts at the junction of the cystic duct, common bile duct, and common hepatic duct, by integrating the patency or obliteration of the common hepatic duct into a single classification.

Applying these classifications to cases with cystic dilations was particularly challenging in this study. Differentiating between types like IIB and III based solely on intraoperative morphological data proved difficult. For example, during trans-cystic cholangiography (when the cystic duct is patent), contrast retention within the cyst may reflect complete or partial choledochal atresia but does not allow visualization of the hilum and intrahepatic ducts if the common hepatic duct is atritic or fibrotic only in its distal portion. In specific cases, preoperative studies such as hepatobiliary scintigraphy were also considered, evaluating bile flow from the parenchyma to the intrahepatic ducts and its passage through the hilum. We extend this as a recommendation to complement preoperative imaging studies upstream (from the liver to its hilum) with intraoperative assessments downstream (from the gallbladder through all proximal and distal extrahepatic ducts). These approaches could 1) more accurately characterize conventional atresia, and 2) describe the extent of atresia affecting the cyst-isolating segments.

Table 2: New Classification for Cystic Biliary Atresia.

Cystic Biliary Atresia Classification (CBA)	
Type - by cyst location	
CBA 1	Cyst located above the cystic duct
CBA 2	Cyst in contact with the confluence of a patent cystic duct
CBA 3	Cyst in contact with the confluence of a non-patent cystic duct
CBA 4	Cyst located below the cystic duct
Subtype - cyst communication	
a	Cyst with proximal communication with the biliary tract.
b	Isolated cyst without proximal or distal communication with the biliary tract.

Cystic Biliary Atresia Classification (CBA)								
Type	**CBA 1**		**CBA 2**		**CBA 3**		**CBA 4**	
Subtype	a	b	a	b	a	b	a	b
No. pacientes	3	7	4	12	1	1	1	3

Frequency distribution of cases in each type and subtype.

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Based on the morphological characteristics of the cystic dilations observed, we propose a new classification specific to cystic biliary atresia, aiming to simplify the description of patients and intraoperative findings based on extrahepatic biliary anatomy. (Table 2) outlines this classification and the patient distribution in this review. (Figure 2) illustrates the two primary criteria used to define this new typology: the location of the dilation concerning the cystic duct, the patency of the cystic duct, and the configuration of bile flow communication with the cyst. It is worth noting that in subtypes such as "b," cholangiographic findings alone may be insufficient. Unlike the original Kasai classification, this new classification integrates surgical dissection findings, cholangiography, and novel intraoperative techniques, such as indocyanine green staining, to highlight biliary pathways downstream from the liver.

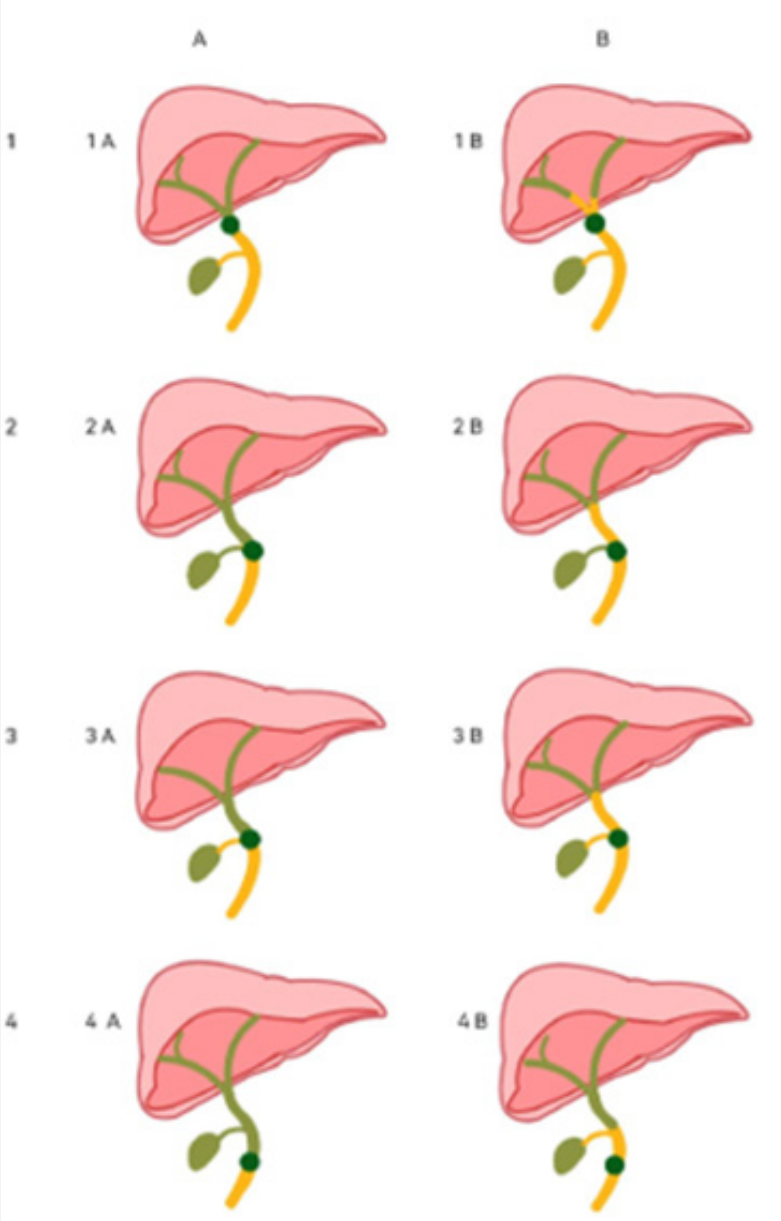

Figure 2: Diagram illustrating the hepatobiliary anatomical considerations of CBA. Diagram illustrating the anatomical considerations of CBA. Dark green dot = represents the cyst and indicates its location within the bile duct. Light green = represents the portion of the bile duct that is likely patent or has potential for flow (depending on the atresia). Yellow = represents segments with obliteration or probable absence of bile flow.

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It is noteworthy that the frequency distribution of patients in CBA was significantly concentrated in subtype 2b, as this anatomical variation specifically complicates alignment with the original and modified Kasai classifications and the anatomical features we identified. It could be argued that the "a" subtypes of the new classification are fully explained by a distal obliterative process, which in most cases likely affects the common bile duct (Kasai I) (CBA 1, 2, or 3) or the common hepatic duct (Kasai IIa) (CBA 4). Both cases are easily distinguishable once patency is confirmed, and the cyst's location is identified.

When observing a non-communicating cystic dilation, neither proximal nor distal to the midpoint of the biliary tract (between the hepatic and common bile ducts), the two apparent possibilities would be Kasai IIb or III atresias. However, this assumption is challenged due to the patency of the cystic duct, which, as described in other studies, is affected in these two forms of atresia. These atresias, theoretically of vertical extension, raise the question of whether they should be considered multiple atretic processes. It could be hypothesized that CBA 3b is more associated with Kasai III due to a more extensive obliterative process involving the extrahepatic biliary tract, likely related to congenital anomalies (given the involvement of the porta hepatis). On the other hand, CBA 2b may be more consistent with Kasai IIb due to the absence of involvement of the hepatic confluences and, consequently, possibly sparing the cystic duct in some cases. (Table 3) summarizes the patients classified as CBA 2b, grouped according to their previously assigned Kasai classification. To fully validate the findings of this study, it would be important to prospectively apply this classification to a larger cohort of neonates with cystic biliary atresia. This would enable consideration of the morphological and functional distributions described in this study from the initial clinical assessment of the patient.

Table 3: Number of patients classified as 'CBA 2b' grouped according to Kasai's classification.

No.	Author	Classification (CBA)	Kasai Classification	Modified Kasai Classification
1	[16]	CBA 2b	Type III	Type IIIb1
2	[17]		Type III	Type IIIb1
3	[20]		Type III	Type IIIb1
4			Type III	Type IIIb1
5	[21]		Type III	Type IIIb1
6			Type III	Type IIIb1
7	[25]		Type IIb	Type IIb1
8			Type III*	Type IIIb1
9	[26]		Type III	Type IIIb1
10	[30]		Type III	Type III d γ*
11	[34]		Type IIb	Type IIb1
12	[36]		Type III	Type IIIb1

Conclusion

Extrahepatic cyst-associated biliary atresia represents a complex and rare variant of the disease, posing challenges in both diagnosis and treatment. Despite advances in the Kasai classification, it is evident that these classifications do not fully capture the anatomical and functional variability of cases with cystic dilations, which could influence critical surgical decisions. The prevalence of biliary atresia types in this variant, with a clear predominance of type III, aligns with findings in previous studies but underscores the need for a more flexible and detailed classification to properly address differences in cyst characteristics, their connection to the biliary system, and their functionality. Accurate identification and classification of these cases are crucial to avoid misdiagnosis and treatment delays, which in turn could improve prognosis and reduce long-term hepatic complications.

Author Contribution

Forero Escobedo S conceptualized the study, contributed to data analysis and interpretation, drafted the manuscript, and approved the final version. Gonzalez Rodriguez SM contributed to data collection, data analysis, and manuscript drafting. Sanchez Tibaduiza S participated in the development of the methodology, data collection, and critical revision of the manuscript for intellectual content.

References

1. Lakshminarayanan B, Davenport M (2016) Biliary atresia: A comprehensive review. J Autoimmun 73: 1-9.

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DOI: 10.19080/OAJS.2025.16.555942

2. Madadi Sanjani O, Rohrbacher V, Uecker M (2024) Cystic Biliary Atresia a Single-Center, Retrospective Analysis. Dtsch Arztebl Int 121(19): 641-642.

3. Vij M, Rela M (2020) Biliary atresia: pathology, etiology and pathogenesis. Future Sci OA 6(5): 1-16.

4. Shin HJ, Yoon H, Han SJ, Ihn K, Koh H, et al. (2021) Key imaging features for differentiating cystic biliary atresia from choledochal cyst: prenatal ultrasonography and postnatal ultrasonography and MRI. Ultrasonography 40(2): 301-311.

5. Kelly DA, Bucuvalas JC, Alonso EM, Karpen SJ, Allen U, et al. (2013) Long-term medical management of the paediatric patient after liver transplantation: 2013 practice guideline by the American association for the study of liver diseases and the American society of transplantation. Liver Transplantation 19(8): 798-825.

6. Stormon MO, Hardikar W, Evans HM, Hodgkinson P (2020) Paediatric liver transplantation in Australia and New Zealand: 1985-2018. J Paediatr Child Health 56(11): 1739-1746.

7. Baumann U, Karam V, Adam R (2022) Prognosis of Children Undergoing Liver Transplantation: A 30-Year European Study. Paediatrics 150(4).

8. Kasai M (1974) Treatment of biliary atresia with special reference to hepatic Porto-enterostomy and its modifications. Progress in paediatric surgery 6: 50-52.

9. Nakamura H, Murase N, Koga H, Cazares J, Lane GJ, et al. (2016) Classification of biliary atresia in the laparoscopic era. Pediatr Surg Int 32(12): 1209-1212.

10. Davenport M (2012) Biliary atresia: clinical aspects. Semin Pediatr Surg 21(3): 175-84.

11. Brandon G O'Connor DO, Richard B Towbin MD, Carrie M Schaefer MD, Alexander J Towbin MD (2024) Cystic Biliary Atresia. Appl Rad Oncol (5): 3-6.

12. Zhou W, Zhou L (2021) Ultrasound for the Diagnosis of Biliary Atresia: From Conventional Ultrasound to Artificial Intelligence. Diagnostics (Basel) 12(1): 1-14.

13. Page MJ, McKenzie JE, Bossuyt PM, Bourton I, Hoffmann TC et al. (2021) The PRISMA 2020 statement: An updated guideline for reporting systematic reviews. BMJ 372(71): 1-9.

14. Tsuchida Y, Kawarasaki H, Iwanaka T, Uchida H, Nakanishi H, et al. (1995) Antenatal diagnosis of biliary atresia (type I cyst) at 19 weeks' gestation: differential diagnosis and etiologic implications. J Pediatr Surg 30(5): 697-699.

15. Matsubara H, Oya N, Suzuki Y, Kajiura S, Suzumori K, et al. (1997) Is it possible to differentiate between choledochal cyst and congenital biliary atresia (type I cyst) by antenatal ultrasonography? Fetal Diagn Ther 12(5): 306-308.

16. Takahashi A, Tsuchida Y, Hatakeyama S, Suzuki N, Kuroiwa M, et al. (1997) A peculiar form of multiple cystic dilatations of the intrahepatic biliary system found in a patient with biliary atresia. J Pediatr Surg 32(12): 1776-1779.

17. Iwai N, Deguchi E, Sasaki Y, Idoguchi K, Yanagihara J (1999) Antenatal diagnosis of biliary atresia (noncorrect able cyst type): a case report. Eur J Pediatr Surg 9(5): 340-342.

18. Shiono S, Yamagiwa I, Obata K, Okuyama N, Ouchi T, et al. (2000) A case of biliary atresia with cystic dilatation of the extrahepatic bile duct and polysplenia syndrome. Pediatr Surg Int 16(8): 599-601.

19. Casaccia G, Bilancioni E, Nahom A, Trucchi A, Aite L, et al. (2002) Cystic anomalies of biliary tree in the foetus: is it possible to make a more specific prenatal diagnosis? J Pediatr Surg 37(8): 1191-1194.

20. De Matos V, Erlichman J, Russo PA, Haber BA (2005) Does "cystic" biliary atresia represent a distinct clinical and etiological subgroup? A series of three cases. Pediatr Dev Pathol 8(6): 725-731.

21. Sookpotarom P, Vejchapipat P (2007) Non-correctable biliary atresia with large extrahepatic cyst: a report of two cases. Eur J Pediatr Surg 17(4): 295-297.

22. Obaidah A, Dhende NP, Mane SB, Acharya H (2009) Biliary atresia associated with choledochal cyst. Afr J Paediatr Surg 6(1): 61-62.

23. Nori M, Venkateshwarlu J, Vijay Sekhar, Prasad GR (2013) Extrahepatic biliary atresia with choledochal cyst: Prenatal MRI predicted and post natally confirmed: A case report. Indian J Radiol Imaging 23(3): 238-242.

24. Adewole VA, Wright NJ, Hallows R, Davenport M (2014) Antenatally detected cystic biliary atresia: differential diagnoses of a double bubble. Springer plus 19(3): 1-5.

25. Parra D, Fecteau A, Daneman A (2016) Findings in percutaneous cholangiography in two cases of Type III cystic biliary atresia (with ultrasound correlation). BJR Case Rep 2(2): 1-3.

26. Schooler GR, Mavis A (2018) Cystic biliary atresia: A distinct clinical entity that may mimic choledochal cyst. Radiol Case Rep 13(2): 415-418.

27. Sakamoto N, Muraji T, Masumoto K, Yanai T, Ono K (2018) Intrahepatic biliary cyst after a Kasai procedure. J Pediatr Surg Case Rep 31: 32-35.

28. Koshinaga T, Ohashi K, Ono K, Kaneda H, Furuya T (2018) Obliterative cholangiopathy in acquired cystic biliary atresia type III after cyst perforation: a case report. BMC Pediatr 18(1): 1-7.

29. Azna AA, Hayati F, Azizan N, Che Noh CI, Osman M (2018) Cystic biliary atresia: A delayed diagnosis of obstructive jaundice. Turkiye Klinikleri J Case Rep 26(2): 102-104.

30. Kimura T, Hirabayashi T, Ishido K, Kobayashi T, Saito T, et al. (2018) Biliary atresia with cystic dilatation of the common bile duct. J Pediatr Surg Case Rep 38: 16-18.

31. Masui D, Fukahori S, Mizuochi T, Watanabe Y, Fukui K, et al. (2019) Cystic biliary atresia with paucity of bile ducts and gene mutation in KDM6A: a case report. Surg Case Rep 5(1): 1-6.

32. Nandan R, Jana M, Yadav DK, Goel P (2021) Cystic biliary atresia with congenital absence of portal vein. BMJ Case Rep 14(1): 1-4.

33. Berkowitz CL, Peters AW, Stratigis JD, Barone PD, Kadenhe Chiweshe AV, et al. (2021) Cystic biliary anomaly in a newborn with features of choledochal cyst and cystic biliary atresia. J Pediatr Surg Case Rep 66: 1-4.

34. Prata R, Nunes A, Borges C, Soares E (2021) Cystic biliary atresia: an uncommon variant of biliary atresia in which a gallbladder may be present. Eurorad Case Pp: 1-8.

35. Kunwar S, BC B, Sah RK (2021) Biliary Atresia with Extrahepatic Cyst: A Diagnostic Dilemma. Cureus 13(8): 1-6.

36. Reuland C, Cappiello CD (2021) Cystic biliary atresia masquerading as a choledochal cyst. J Pediatr Surg Case Rep 74: 1-5.

37. Rahamtalla D, Al Rawahi Y, Jawa ZM, Wali Y (2022) Cystic biliary atresia in a neonate with antenatally detected abdominal cyst. BMJ Case Rep 15(2): 1-3.

38. Lee NW, Karim NKA, Singh PSD (2023) A rare case of cystic biliary atresia. Malays J Med Health Sci 19(3): 380-382.

39. Wang M, Liao J, Li S, Zhang S, Huang J (2023) Laparoscopic indocyanine green fluorescence imaging technique in rare type II cystic biliary atresia. Photodiagnosis Photodyn Ther Pp: 44.

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DOI: 10.19080/OAJS.2025.16.555942

40. Issenman J, Martin K (2025) Common bile duct cysts in neonates, not always a choledochal cyst: A case report. J Pediatr Surg Case Rep 112: 1-5.

41. Yamataka A, Ohshiro K, Okada Y, Hosoda Y, Fujiwara T, et al. (1997) Complications after cyst excision with hepatic enterostomy for choledochal cysts and their surgical management in children versus adults. J Pediatr Surg 32(7): 1097-1102.

42. Sasaki H, Nio M, Ando H, Kitagawa H, Kubota M, et al. (2021) Anatomical patterns of biliary atresia including hepatic radicles at the porta hepatis influence short- and long-term prognoses. Journal of hepato-biliary-pancreatic sciences 28(11): 931-941.

43. Fischler B, Papadogiannakis N, Nemeth A (2001) Aetiological factors in neonatal cholestasis. Acta paediatric (Oslo, Norway: 1992) 90(1): 88-92.

44. Lobeck IN, Sheridan R, Lovell M, Dupree P, Tiao GM, et al. (2017) Cystic Biliary Atresia and Choledochal Cysts Are Distinct Histopathologic Entities. The American journal of surgical pathology 41(3): 354-364.

45. Hartley JL, Davenport M, Kelly DA (2009) Biliary atresia. Lancet 374(9702): 1704-1713.

46. Asai A, Wu JF, Wang KS, Yamataka A, Nio M, et al. (2022) The Outcome of Patients with Cystic Biliary Atresia with Intact Proximal Hepatic Ducts Following Hepatic-Cyst-Jejunostomy. Journal of paediatric gastroenterology and nutrition 75(2): 131-137.

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