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Caroli Disease and Caroli Syndrome Mimicking Cystic Biliary Atresia in Infancy: Age-Associated Presentations and Surgical Implications

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Abstract

Background/Purpose.

Caroli-spectrum disorders presenting in infancy may mimic cystic biliary atresia (CBA). We examined age-associated presentations and features distinguishing infantile Caroli-spectrum disease from CBA.

Methods.

We retrospectively reviewed 15 children with Caroli disease/syndrome and 21 infants with CBA managed from 2013 through March 2026. Caroli patients were divided into two age groups: younger than 1 year and 1 year or older. Infantile Caroli cases were compared with CBA using Mann-Whitney U and Fisher exact tests; analyses were descriptive.

Results.

Caroli-spectrum disease presented from 23 days to 12.5 years (median, 5.8 years). Infants had predominantly cholestatic presentations, whereas older children more often had portal hypertension related bleeding. Conjugated bilirubin was 108.3 [80.3–119.0] μmol/L in infants and 4.3 [3.4–5.8] μmol/L in older children. Compared with CBA, infantile Caroli cases showed substantial biochemical overlap (all $p >= 0.13$). Gallbladder nonvisualization or small size occurred in both groups and was not independently discriminatory. Cystic/saccular intrahepatic duct dilatation was documented in 4/4 infantile Caroli cases and 0/16 assessable CBA cases ($p < 0.001$). CBA management included eight Kasai portoenterostomies and one primary liver transplantation.

Conclusions.

Infantile Caroli-spectrum disease may overlap substantially with CBA clinically and biochemically. In our cohort, cystic or saccular involvement of the intrahepatic biliary tree was an observation that raised suspicion for Caroli-spectrum disease and prompted careful reassessment of the biliary anatomy. This finding should not be interpreted as a stand-alone diagnostic criterion.

Introduction

Caroli disease is characterized by nonobstructive segmental, saccular, or fusiform dilatation of the intrahepatic bile ducts. When this biliary abnormality is accompanied by congenital hepatic fibrosis, portal hypertension (PH), and/or the renal phenotype of autosomal-recessive polycystic kidney disease (ARPKD), the term Caroli syndrome is commonly used [1-5]. These entities belong to the ductal-plate malformation spectrum and may present at any age with cholestasis, recurrent cholangitis, hepatolithiasis, PH related bleeding, or complications of associated renal disease. Cystic biliary atresia

(CBA) is a less common morphologic subtype of biliary atresia in which a cystic lesion is present within an otherwise obliterated extrahepatic biliary tree, usually at or near the porta hepatis [6-9]. The distinction is surgically consequential. CBA requires timely assessment for Kasai portoenterostomy, whereas a portoenterostomy does not correct the diffuse intrahepatic ductal abnormality or congenital hepatic fibrosis of Caroli-spectrum disease. Conversely, excessive diagnostic delay in a child with true CBA may reduce the opportunity for effective native-liver drainage.

The clinical problem is most difficult in infancy. A child with Caroli-spectrum disease may have jaundice, acholic stool, conjugated hyperbilirubinemia, elevated cholestatic enzymes, and a cystic structure in the liver or porta hepatis. These findings can be interpreted as CBA or a choledochal cyst unless the intrahepatic biliary tree is examined deliberately. The question underlying the present study arose from our referral experience with infants in whom CBA or choledochal cyst had initially been suspected and a Kasai procedure had been considered or attempted before reassessment at our center. We therefore reviewed our pediatric Caroli-spectrum cohort with two objectives: first, to characterize how the clinical presentation differed between infancy and later childhood; and second, to compare infantile Caroli-spectrum cases with children classified as CBA, focusing on the practical clinical, biochemical, and anatomic observations that informed treatment. The study was not designed to validate a stand-alone diagnostic test. It describes a clinical approach developed through longitudinal pediatric hepatobiliary practice.

Methods. This retrospective observational comparative study was conducted in the Department of Pediatric Surgery of the Republican Specialized Scientific-Practical Medical Center of Pediatrics, Tashkent, Uzbekistan. The center is a national tertiary referral institution receiving children from regions throughout Uzbekistan and from neighboring countries. Records from 2013 through March 2026 were reviewed. Children younger than 18 years managed during the study period for Caroli disease/syndrome, or CBA were eligible. Fifteen children with Caroli-spectrum disease and 21 children classified as CBA were included. No patient was excluded after cohort identification because the purpose was to describe the complete available institutional experience; however, the denominators for individual variables differ because some historical records were incomplete.

Caroli disease was defined clinically and radiologically by segmental, multifocal, or diffuse cystic/saccular dilatation of the intrahepatic bile ducts. Caroli syndrome was used when this ductal abnormality coexisted with evidence of congenital hepatic fibrosis, PH, and/or an associated fibropolycystic renal phenotype. For analyses encompassing both diagnoses, the term Caroli-spectrum disorder was used.

CBA was diagnosed from the combined clinical, biochemical, ultrasonographic, cross-sectional imaging, operative, and histopathologic findings available in routine care. Ultrasonography was available for all children. MRI/MRCP was performed in 14/15 Caroli-spectrum patients and 6/21 CBA patients; CT or CT angiography was used selectively. Nine CBA patients underwent surgery, including eight Kasai portoenterostomies and one primary liver transplantation, providing operative confirmation of the

diagnosis. Intraoperative cholangiography was performed selectively in the operated CBA subgroup, while histopathologic examination was available in eight patients. Because cholangiography was used only in selected CBA cases and was not applicable to the Caroli-spectrum cohort, its findings were not included in the comparative analysis. The remaining 12 CBA cases were classified from the consistent clinical-radiologic phenotype and were retained to represent the real-world referral population. In the Caroli cohort, five children underwent surgery and histopathology was available in two.

Within the Caroli-spectrum cohort, associated ARPKD was assessed by renal ultrasonography and clinical evaluation. Genetic testing was not performed routinely in all children presenting with cholestasis. Genetic confirmation was available in three of the four Caroli-spectrum patients diagnosed with ARPKD; the remaining patient was diagnosed on the basis of the clinical and radiologic findings.

The following variables were extracted: demographic characteristics, including sex, date of birth, and age at first specialist admission; presenting manifestations, including jaundice, acholic stool, bleeding, cholangitis, and other disease-related complications; biochemical markers of cholestasis, including serum alkaline phosphatase (ALP), gamma-glutamyl transferase (γGT), and total and conjugated bilirubin; markers of hepatocellular injury, including alanine aminotransferase (ALT) and aspartate aminotransferase (AST); liver synthetic and coagulation parameters, including serum albumin, international normalized ratio (INR), and activated partial thromboplastin time; associated renal disease; ultrasonographic and MRI/MRCP characteristics of the gallbladder, intrahepatic bile ducts, and cystic lesions; liver stiffness measurements obtained by 2D shear wave elastography (2D SWE); operative procedures; and histopathologic findings, when available. Laboratory values used for comparison were those obtained at the index admission before the relevant intervention. Imaging variables were abstracted from the contemporaneous examinations and reports used in clinical decision-making. Particular attention was paid to the location of the cystic abnormality, the presence of diffuse or segmental intrahepatic duct dilatation, and whether the intrahepatic abnormality was cystic/saccular rather than mild nonspecific ductal prominence. Imaging was not reread in a blinded central review because the objective was to describe the diagnostic reasoning used in routine practice.

The Caroli-spectrum cohort was divided into infants younger than 1 year (n=4) and children aged 1 year or older (n=11). This analysis assessed age-associated patterns within the Caroli cohort. Because the CBA cohort consisted entirely of infants, the comparison between diagnoses was restricted to the four infantile Caroli-spectrum patients and the 21 CBA patients. The fourth infantile Caroli patient, aged 349 days, was retained because the predefined age threshold was younger than 1 year; individual patient data are presented to show that this child was anicteric and biochemically different from the three younger cholestatic infants.

Continuous variables were summarized as median [interquartile range], and categorical variables as n/N. Comparisons between independent groups were performed using the two-sided Mann–Whitney U test for continuous variables and the two-sided Fisher exact test for categorical variables. Missing values were not imputed; analyses were based on available observations, and the corresponding denominators

are reported. A nominal p value <0.05 was used as a reference threshold. Given the small infantile Caroli-spectrum subgroup (n=4) and the number of variables examined, all between-diagnosis analyses were considered exploratory. Non-significant results were not interpreted as evidence of equivalence. No adjustment for multiple comparisons was applied, and p values were interpreted in conjunction with the observed distributions and clinical findings. Analyses were performed in Python using SciPy.

The study protocol was approved by the Ethics Committee of the Republican Specialized Scientific-Practical Medical Center of Pediatrics, Tashkent, Uzbekistan (IRB approval No. 2026-001; March 25, 2026). Given the retrospective design of the study and the use of anonymized (de-identified) clinical data, the requirement for informed consent was waived by the Ethics Committee. The study was conducted in accordance with the ethical principles of the Declaration of Helsinki and its subsequent amendments.

Results. Fifteen children with Caroli-spectrum disease were included: eight girls and seven boys. Fourteen were classified clinically as Caroli syndrome and one as Caroli disease. Age at first admission ranged from 23 days to 12.5 years, with a median of 2105 days (5.8 years) [680-3359 days]. Four children presented during the first year of life and 11 at 1 year of age or older. ARPKD was present in four children (27%), all in the older group (Table 1)

Table 1. Characteristics of the pediatric Caroli-spectrum cohort by age group

Variable	Infants <1 year (n=4)	Children >=1 year (n=11)	p value
Male sex, n/N	3/4	4/11	0.282
Age at presentation, days	140.5 [82.3-221.5]	2713 [2046.5-3497.5]	0.001
Jaundice, n/N assessable	3/3	3/10	0.070
Acholic stool, n/N assessable	2/2	0/10	0.015
Any bleeding manifestation, n/N	0/4	7/11	0.077
ARPKD, n/N	0/4	4/11	0.516
ALP, U/L	884.6 [243.6-1588.5]	518 [293-734] (n=9)	0.710
GGT, U/L	165.8 [102.0-210.3]	35 [20-58] (n=9)	0.353
ALT, U/L	160.5 [110.6-193.5]	30 [25.5-55.5] (n=10)	0.039
AST, U/L	153.3 [132.7-222.8]	32 [25.8-89.3] (n=10)	0.028
Total bilirubin, µmol/L	248.6 [178.6-277.4]	17.7 [15.9-19.3] (n=10)	0.177
Conjugated bilirubin, µmol/L	108.3 [80.3-119.0]	4.3 [3.4-5.8] (n=10)	0.077
Albumin, g/L	38.0 [33.3-41.3]	30.0 [29.5-38.5]	0.512
INR	1.12 [1.01-1.51]	1.16 [1.07-1.34] (n=9)	0.825
2D-SWE, kPa	48.6 [8.9-96.2]	19.2 [14.6-23.4] (n=4)	1.000

Values are median [IQR] unless otherwise indicated. Denominators reflect available data. ARPKD, autosomal-recessive polycystic kidney disease; 2D-SWE, two-dimensional shear-wave elastography.

The infantile presentation was predominantly cholestatic. Jaundice was documented in 3/3 infants with available data and acholic stool in 2/2. Median conjugated bilirubin was 108.3 [80.3-119.0] µmol/L in infants compared with 4.3 [3.4-5.8] µmol/L in older children. ALT and AST were also higher in infants (p=0.039 and p=0.028, respectively). The conjugated-bilirubin difference did not reach conventional statistical significance (p=0.077), reflecting the very small infant subgroup and the inclusion of the 349-day-old anicteric child.

Older children presented more commonly with manifestations associated with established PH or cholangitis. Bleeding manifestations were recorded in 7/11 older children and in none of the four infants; five had gastrointestinal bleeding and two had epistaxis in the setting of chronic liver disease. Recurrent cholangitis was the principal presentation in two older children. All four ARPKD-associated cases occurred in the older group. These findings represent cross-sectional age-associated differences in presentation rather than proof of a uniform within-patient sequence.

Three children, aged 23, 102, and 179 days, had marked conjugated hyperbilirubinemia and clinical cholestasis. The fourth child was evaluated at 349 days, had a normal conjugated bilirubin concentration of 3.5 μmol/L. Despite the difference in biochemical presentation, cystic, mixed, or ectatic intrahepatic duct abnormalities were documented in all four children on ultrasonography and/or MRI/MRCP (Fig. 1). None had ARPKD at the time of infantile evaluation.

The four infantile Caroli-spectrum patients and the 21 CBA patients occupied an overlapping age range (median, 140.5 [82.3-221.5] versus 71 [65-132] days; p=0.394). Laboratory values also overlapped substantially (Table 2). Median conjugated bilirubin was 108.3 [80.3-119.0] μmol/L in infantile Caroli-spectrum disease and 129.3 [112.7-159.0] μmol/L in CBA (p=0.132). There was no clear separation by ALP, GGT, ALT, AST, total bilirubin, albumin, INR, or liver stiffness according to 2D SWE (all p>=0.13)

Table 2. Exploratory comparison of infantile Caroli-spectrum disease and cystic biliary atresia

Variable	Infantile Caroli spectrum (n=4)	Cystic biliary atresia (n=21)	p value
Age at presentation, days	140.5 [82.3-221.5]	71 [65-132]	0.394
Male sex, n/N	3/4	6/21	0.116
ALP, U/L	884.6 [243.6-1588.5]	812 [489-1068.8] (n=19)	0.845
GGT, U/L	165.8 [102.0-210.3]	199 [135.9-735] (n=18)	0.342
ALT, U/L	160.5 [110.6-193.5]	160 [119-245.5] (n=20)	0.737
AST, U/L	153.3 [132.7-222.8]	197 [150.5-336] (n=20)	0.416
Total bilirubin, μmol/L	248.6 [178.6-277.4]	214 [194.9-266.4]	0.803
Conjugated bilirubin, μmol/L	108.3 [80.3-119.0]	129.3 [112.7-159.0]	0.132
Albumin, g/L	38.0 [33.3-41.3]	32 [30-39] (n=17)	0.560
INR	1.12 [1.01-1.51]	1.23 [1.05-1.36] (n=14)	0.798
2D-SWE, kPa	48.6 [8.9-96.2]	13.2 [11.7-22.6] (n=8)	0.799
Cystic/saccular intrahepatic duct dilatation, n/N assessable	4/4	0/16	<0.001

Values are median [IQR] unless otherwise indicated. Comparisons are exploratory.

Gallbladder findings also overlapped between the two groups. Among infantile Caroli-spectrum patients with evaluable native gallbladder findings, the gallbladder was nonvisualized in one child and visualized but small in two. MRI described the gallbladder as atretic in two patients, although ultrasonography identified a small gallbladder in one of them. Among 16 CBA patients with available ultrasonographic gallbladder data, the gallbladder was nonvisualized in six and measurable in ten. Thus, gallbladder nonvisualization or reduced size was not specific to either condition and could not independently distinguish CBA from infantile Caroli-spectrum disease.

The most useful practical distinction was the pattern and location of the biliary abnormality. All four infantile Caroli-spectrum patients had a cystic/saccular, mixed, or ectatic abnormality involving the intrahepatic biliary tree. No CBA patient had a cystic or saccular intrahepatic duct pattern. Thus, cystic/saccular intrahepatic duct dilatation was present in 4/4 infantile Caroli-spectrum patients and 0/16 assessable CBA patients (Fisher exact $p<0.001$). The CBA cystic lesion was instead hilar or extrahepatic. (Fig. 2)

Liver stiffness measurements were available in a limited number of patients and showed substantial variability. In the infant comparison, 2D shear-wave elastography values overlapped between Caroli-spectrum disease and CBA ($p=0.799$). Because of the small sample size, the influence of cholestasis, and the use of different elastography techniques across age groups, liver stiffness was not considered useful for differential diagnosis.

Five patients with Caroli-spectrum disease had undergone surgical procedures. Three operations were performed at our center for complications of portal hypertension: Hassab devascularization in two patients and a Sugiura procedure in one. Before referral to our center, two patients with Caroli-spectrum disease had undergone bilioenteric reconstruction at outside institutions under a presumed alternative diagnosis of a cystic biliary abnormality. Both subsequently developed recurrent or persistent cholangitis and remained under medical management for ongoing biliary infection at the time of the present analysis. Among the 21 patients classified as having CBA, nine underwent operative treatment: eight Kasai portoenterostomies and one primary liver transplantation. Histopathologic examination was available in eight patients. Several of the remaining patients presented late, with advanced liver disease, and were managed along a pathway directed toward assessment for primary liver transplantation rather than portoenterostomy.

Discussion

This study describes two clinically distinct modes of presentation within a pediatric Caroli-spectrum cohort and relates them to a specific surgical diagnostic problem. Infants predominantly presented with cholestasis, whereas older children more often came to specialist attention because of PH related bleeding or recurrent cholangitis. When the infantile Caroli cases were compared with CBA, routine laboratory tests showed extensive overlap. The most useful observation in our practice was not a biochemical threshold but the anatomic pattern of the biliary tree: communicating cystic, saccular,

mixed, or ectatic intrahepatic duct disease favored the Caroli spectrum, whereas the CBA lesion was centered at the hilum or extrahepatic remnant without cystic intrahepatic duct dilatation.

The distinction matters because the therapeutic pathways are fundamentally different. CBA is a time-sensitive form of biliary atresia in which early bile drainage may preserve native-liver survival [6-8, 10, 11]. Caroli-spectrum disease is an intrahepatic ductal-plate malformation; treatment depends on its distribution and dominant complication and may range from observation and treatment of cholangitis to PH surgery, hepatic resection for localized disease, or transplantation in advanced diffuse disease. A Kasai portoenterostomy cannot correct the underlying intrahepatic abnormality. It therefore exposes a misclassified child to an unnecessary major operation, adhesions, and altered hilar anatomy, while delay caused by excessive investigation is equally undesirable in genuine CBA.

The consequences of diagnostic uncertainty were evident in our referral experience. Two children with Caroli-spectrum disease had undergone bilioenteric reconstruction before referral under an alternative presumed diagnosis and subsequently developed persistent cholangitis. These observations do not allow conclusions regarding the appropriateness of the initial procedures, because complete preoperative information from the referring institutions was not available. However, they illustrate the potential clinical consequences of undertaking biliary reconstruction without fully defining whether the cystic abnormality represents an extrahepatic lesion or communicating intrahepatic duct disease.

Our practical approach begins with a deliberate question: is the apparent cyst an isolated hilar/extrahepatic structure within an atretic biliary tree, or does it represent abnormal intrahepatic ducts? On ultrasonography, we examine the right and left intrahepatic systems separately, the relationship of the cystic spaces to portal structures, gallbladder morphology, and the presence of a distal duct. MRI/MRCP is used when ultrasonography does not define the anatomy adequately and when the child can undergo the study without unacceptable delay. We also examine the kidneys and search for features of ARPKD or another fibropolycystic phenotype. The absence of renal disease does not exclude Caroli-spectrum disease, as none of our four infantile patients had ARPKD.

Gallbladder morphology should be interpreted with caution. In our infantile Caroli-spectrum cohort, the gallbladder was atretic or nonvisualized in 2 of 4 patients and remained visible, although small or measurable, in the other 2. Similarly, among children classified as having CBA, a measurable gallbladder was present in 11 of 21 patients, whereas it was nonvisualized or atretic in 6 and not assessed in 4. These findings demonstrate substantial overlap in gallbladder appearance between the two conditions. Gallbladder morphology therefore contributed to the overall diagnostic assessment but was not considered sufficiently specific to guide surgical decision-making in isolation. Greater emphasis was placed on the location and configuration of the cystic abnormality and on whether it represented communicating intrahepatic duct dilatation or an isolated hilar or extrahepatic lesion.

Published imaging studies have largely concentrated on differentiating CBA from choledochal cyst rather than from Caroli-spectrum disease. Zhou and colleagues found no intrahepatic bile-duct dilatation in 12 infants with CBA, whereas it was present in most children with choledochal cyst [9]. This supports the

importance of examining the intrahepatic ducts, although the comparison is not identical to ours. CBA and atretic choledochal cyst may also exist on a morphologic continuum, and difficult cases may remain ambiguous despite high-quality imaging [12]. Our data should therefore not be read as establishing an absolute exclusion rule. Rather, they show why a cystic intrahepatic pattern should prompt reconsideration of a diagnosis of CBA before irreversible surgery. The age-associated findings are also clinically informative. The severe cholestatic profile in three young infants contrasted with the near-normal conjugated bilirubin values in most older children, in whom PH dominated. Congenital hepatic fibrosis and ARPKD are well recognized to produce a variable pediatric phenotype, including PH with relatively preserved hepatocellular function [4, 5, 13, 14]. Our cross-sectional data are consistent with this spectrum, but they do not prove that every infant with Caroli disease will later follow the same sequence. Referral patterns, genotype, distribution of ductal disease, infection, and survival may all contribute to the differences between age groups.

Liver stiffness did not distinguish infantile Caroli-spectrum disease from CBA, likely because both cholestasis and fibrosis may increase stiffness. Its role in this setting therefore appears limited to assessment of liver involvement rather than differential diagnosis.

The principal strength of this study is that it reflects a complete real-world pediatric hepatobiliary referral experience with a rare disease and links the observed phenotype directly to surgical decision-making. Individual infant data are presented rather than hidden behind group averages. The study also avoids comparing the entire older Caroli cohort with an exclusively infantile disease and explicitly shows the clinical outlier at 349 days.

Several limitations should be acknowledged. The study was retrospective, single-center, and small, especially the infantile Caroli subgroup. The comparisons are exploratory and cannot establish diagnostic accuracy or biochemical equivalence. Diagnostic evaluation was clinically driven and therefore heterogeneous; MRI/MRCP was used more frequently in Caroli-spectrum disease than in CBA, and imaging was not subjected to blinded central rereview. Caroli diagnoses were usually clinical-radiologic rather than histologic. Nine CBA cases had operative confirmation, whereas 12 were classified from the clinical-radiologic phenotype without operative confirmation, leaving some risk of misclassification. Intraoperative cholangiography was not performed. Missing data reduced the denominators for several symptoms and laboratory variables. Two features warrant particular caution in interpreting the intrahepatic-duct finding. First, the diagnostic labels and the discriminating anatomic feature were not fully independent: a child with cystic or saccular intrahepatic ductal dilatation may have been assigned to the Caroli spectrum rather than to CBA at initial classification, so the observed separation of intrahepatic patterns partly reflects the diagnostic reasoning under study rather than an externally validated criterion. Second, imaging intensity differed between cohorts, with MRI/MRCP obtained in 14 of 15 Caroli-spectrum patients but only 6 of 21 CBA patients, and assessment of the intrahepatic ducts in CBA relying predominantly on ultrasonography; the contrast between cystic intrahepatic dilatation in 4 of 4 infantile Caroli patients and 0 of 16 assessable CBA patients may

therefore partly reflect differential imaging modality and completeness rather than anatomy alone. Finally, the cross-sectional age comparison cannot establish the natural history of an individual patient.

Despite these limitations, the clinical message is practical. A cholestatic infant with a biliary cyst should not be assigned automatically to a Kasai or choledochal-cyst pathway. The intrahepatic ducts must be interrogated deliberately, and the renal phenotype should be assessed. At the same time, this evaluation should be organized rapidly so that a child with true CBA is not disadvantaged by delay.

Conclusions

Pediatric Caroli-spectrum disorders showed age-associated heterogeneity in presentation, with cholestasis predominating in infancy and manifestations of portal hypertension occurring more commonly in older children. Infantile Caroli-spectrum disease overlapped substantially with CBA clinically and biochemically. In our experience, cystic or saccular abnormalities involving the intrahepatic biliary tree represented an important observation that prompted reconsideration of a presumed CBA diagnosis and more detailed assessment of the biliary anatomy. Because diagnostic classification and imaging findings were not independent and imaging assessment differed between groups, this observation should not be regarded as a validated or stand-alone diagnostic criterion.

Declarations

Author Contribution

Author contributions: R.Y. conceived and designed the study, coordinated and participated in data acquisition, performed the statistical analysis, interpreted the findings, drafted the original manuscript, and critically revised subsequent versions. N.K. performed the radiologic assessment, interpreted the ultrasound and MRCP findings, drafted the imaging-related sections, and critically revised the manuscript for important intellectual content. M.A. contributed to the study design and clinical supervision, interpreted the findings, and critically revised the manuscript for important intellectual content. G.T. and A.G. contributed to patient management and clinical data acquisition, interpreted the findings, reviewed the relevant literature, and critically revised the manuscript for important intellectual content. All authors reviewed and approved the final version of the manuscript, gave explicit consent for its submission, and agreed to be accountable for all aspects of the work, including ensuring that any questions concerning the accuracy or integrity of the work are appropriately investigated and resolved. During manuscript preparation, the authors used ChatGPT (OpenAI) to assist with language editing, clarity, and conciseness. All AI-assisted output was critically reviewed, verified, and revised by the authors, who take full responsibility for the accuracy, integrity, and originality of the final manuscript.

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Figures

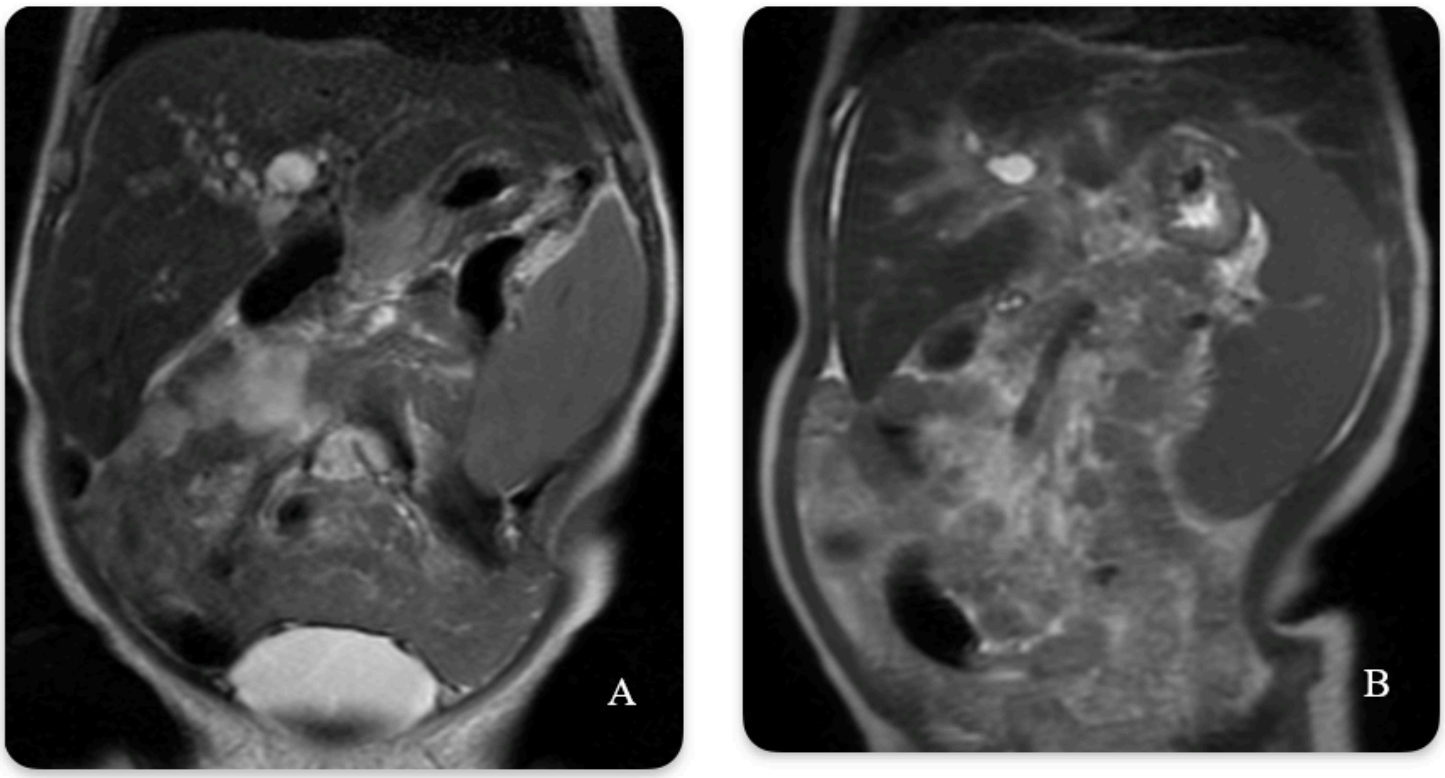


Figure 1

Coronal MR images in two children with Caroli syndrome. **(A)** A dominant well-defined fluid-signal structure and multiple smaller fluid-signal foci are identified along the expected course of the intrahepatic bile ducts, compatible with multifocal saccular ductal ectasia. **(B)** A dominant intrahepatic fluid-signal structure is seen with an additional smaller focus located superiorly. Associated splenomegaly is present. The multiplicity and intrahepatic distribution of the lesions support Caroli-spectrum disease.

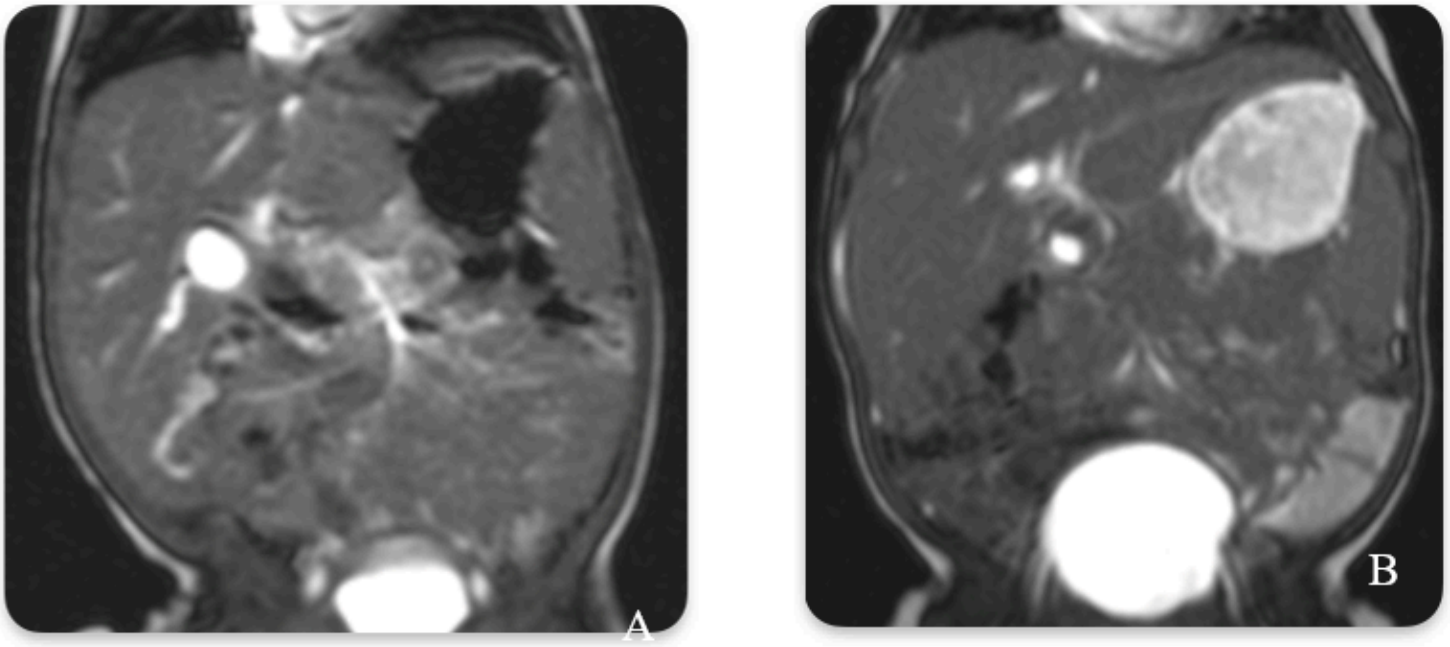


Figure 2

Coronal T2-weighted MR images from two infants with cystic biliary atresia demonstrate variability in the size and location of the cystic component. **(A)**A well-defined, fluid-signal cystic structure is centered at the porta hepatis and is associated with a diminutive, atretic-appearing gallbladder. **(B)**A smaller fluid-signal cystic structure is located within the right hepatic hilum, adjacent to the expected course of the right portal venous branch; the gallbladder is not visualized. Neither case demonstrates multifocal or segmental saccular dilatation of the intrahepatic bile ducts.