

## Differential Diagnosis of Biliary Atresia and Other Causes of Neonatal Cholestasis: A 10-Year Clinical Experience

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**Cite this paper as:** Nematjonov Farrukh Zokirjon ugli, Aliyev Makhmud Muslimovich, Tuychiyev Golibjon Urmonjonovich, Yuldashev Rustam Zafarjanovich, Gofurov Adkham Anvarovich, Abduvaliyeva Chulponoy Mukhammadjonova, Shakirova Roksana, (2025) Differential Diagnosis of Biliary Atresia and Other Causes of Neonatal Cholestasis: A 10-Year Clinical Experience. *Journal of Neonatal Surgery*, 14 (32s), 6298-6305.

### ABSTRACT

#### Introduction

**Biliary atresia (BA)** is a rare but severe fibro-obliterative disease of the bile ducts of unknown etiology, manifesting in the neonatal period. According to various sources, its incidence ranges from 1 in 5,000 to 1 in 22,000 live births [1–7]. BA is the leading cause of surgical (obstructive) cholestasis in infants and represents the most common indication for liver transplantation in pediatric patients [8]. Timely performance of the Kasai procedure (portoenterostomy) — preferably before the age of 60–90 days — significantly improves bile flow, slows the progression of fibrosis, and reduces the likelihood of requiring liver transplantation [9, 10]. However, early diagnosis of BA poses a significant clinical challenge, both in resource-limited settings and in well-developed healthcare systems.

The main difficulty lies in differentiating BA from other causes of neonatal cholestasis — such as idiopathic neonatal hepatitis, progressive familial intrahepatic cholestasis (PFIC), Alagille syndrome, congenital hepatic fibrosis, cystic transformations of the common bile duct, as well as infections (e.g., CMV) and metabolic disorders [10–15]. All these conditions may present with similar clinical features: prolonged jaundice, acholic stools, and hepatomegaly. Among the biochemical markers of cholestasis, elevated levels of direct bilirubin, gamma-glutamyl transferase (GGT), and alkaline phosphatase are particularly important, although they are not specific and must be interpreted in combination with imaging findings [13].

**Abdominal ultrasound (US)** remains the primary method for initial diagnosis [16–18]. It allows evaluation of the shape and size of the gallbladder, identification of the "triangular cord sign" at the hepatic hilum, and the exclusion of other bile duct anomalies. However, the sensitivity and specificity of ultrasound depend on the operator's experience and may be reduced in the early stages of the disease.

Additional diagnostic information can be obtained from magnetic resonance cholangiopancreatography (MRCP) and hepatobiliary scintigraphy, although the availability of these methods is often limited, especially in resource-constrained countries [19]. In recent years, **transient elastography (TE)** has been increasingly introduced as a non-invasive method to assess liver stiffness as an indirect marker of fibrosis severity [20, 21]. Given the rapid progression of fibrosis in BA, TE may serve as a valuable diagnostic tool.

It is assumed that combining ultrasound and elastography improves the accuracy of early detection of BA and allows for timely referral for surgical intervention. However, data on the diagnostic effectiveness of this approach in resource-limited settings are still insufficient. Therefore, the aim of this study is to evaluate the diagnostic value of abdominal ultrasound and transient liver elastography in the early diagnosis of presumed biliary atresia in children with neonatal cholestasis, based on 10 years of clinical experience.

**Objective of the study.** To evaluate the diagnostic criteria for biliary atresia (BA) and other causes of neonatal cholestasis in children based on 10 years of clinical experience.

**Materials and methods:** The retrospective study included 157 neonates and infants with signs of cholestasis who were hospitalized at the Republican Specialized Scientific and Practical Medical Center of Pediatrics (Tashkent) between 2013 and 2023. The diagnostic algorithm included abdominal ultrasound (US), transient elastography (TE) of the liver, and, when indicated, hepatobiliary scintigraphy and MRI with cholangiography. The diagnosis of biliary atresia (BA) was surgically confirmed (via intraoperative cholangiography or liver biopsy) in 43 patients. In 88 other children, BA was considered probable based on a combination of clinical, laboratory, and imaging data. In the remaining patients, other diagnoses unrelated to BA were established using a comprehensive examination, including genetic testing in selected cases.

**Results:** Presumed biliary atresia (BA) was diagnosed in 131 patients (83.4%), of whom 43 (32.8%) had the diagnosis surgically confirmed. The 'triangular cord sign' on ultrasound was detected in 38.3% of cases, and gallbladder abnormalities were found in 65.5%; however, in 35.8% of patients, the gallbladder appeared morphologically normal. The average liver stiffness value measured by transient elastography (TE) in children with presumed BA was significantly higher than in other forms of cholestasis ( $p < 0.05$ ), especially in patients older than 90 days. Among a subset of six patients who underwent scintigraphy, absence of intestinal excretion of the radiopharmaceutical was observed in all cases; BA was confirmed in four of them. MR cholangiography ( $n=12$ ) revealed signs of the cystic form of BA, cystic transformation of the common bile duct, and syndromic BA with polysplenia.

**Conclusion:** The combination of ultrasound examination and transient elastography improves the accuracy of early diagnosis of presumed biliary atresia. However, a comprehensive approach that takes into account laboratory findings and additional imaging methods is crucial for timely differentiation of BA from other causes of cholestasis and for determining the optimal treatment strategy.

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**Keywords:** *biliary atresia, neonatal cholestasis, ultrasound examination, transient elastography*

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## 1. MATERIALS AND METHODS

### *Study Design and Sample Characteristics*

A retrospective study was conducted covering the period from January 2013 to December 2023. The analysis included 157 neonates and infants ( $\leq 6$  months old) with signs of neonatal cholestasis — acholic stools and conjugated hyperbilirubinemia. All patients were hospitalized at the Republican Specialized Scientific and Practical Medical Center of Pediatrics (Tashkent). Initial screening and ultrasound examinations were performed at two regional healthcare institutions, after which the patients were referred to the specialized center for further evaluation and diagnostic verification.

#### Diagnostic Algorithm

All patients underwent standard biochemical testing, including the measurement of alanine aminotransferase (ALT), aspartate aminotransferase (AST), total and direct bilirubin, gamma-glutamyl transferase (GGT), alkaline phosphatase (ALP), albumin, fibrinogen, and international normalized ratio (INR). These parameters were used to assess cytolysis, cholestasis, and the liver's synthetic function. In suspected cases of cytomegalovirus (CMV) infection, serological testing (IgM/IgG) and, when necessary, polymerase chain reaction (PCR) were performed.

Abdominal ultrasound was performed for all patients. Evaluated parameters included the size and morphology of the gallbladder (presence, length, shape, wall), the presence of the "triangular cord" sign in the hepatic hilum, condition of the extrahepatic bile ducts, liver structure, presence of ascites, spleen size, and any anomalies. Signs of portal hypertension (e.g., splenomegaly, collateral circulation) were also considered [18].

Transient elastography (TE) of the liver (FibroScan) was performed selectively in patients without signs of ascites, as the presence of free fluid in the abdominal cavity interferes with accurate tissue stiffness measurements. Measurements were obtained using a specialized pediatric S-probe designed for patients weighing less than 10–15 kg, enabling valid liver stiffness readings in infants. The obtained stiffness values were considered supportive in the diagnostic process and were not interpreted using rigid threshold criteria.

Hepatobiliary scintigraphy with Tc-99m-labeled radiopharmaceuticals was selectively conducted when ultrasound and/or

elastography results were inconclusive. A key diagnostic criterion in favor of BA was the absence of radiotracer visualization in the intestines within 24 hours.

Magnetic resonance imaging (MRI) of the abdominal organs with cholangiography (MRCP) was performed in patients with suspected biliary tract malformations, cystic forms of BA, syndromic variants (e.g., with polysplenia), and vascular anomalies. This method allowed for detailed visualization of the biliary anatomy and associated abnormalities.

In selected cases, depending on the clinical picture, additional investigations were performed, including chest X-rays (to identify vertebral anomalies), echocardiography (for congenital heart defect diagnostics), and genetic testing — particularly in cases of suspected Alagille syndrome or progressive familial intrahepatic cholestasis (PFIC). The availability and clinical appropriateness determined the feasibility of these investigations.

### **Diagnostic Criteria**

- Confirmed biliary atresia (n = 43): Diagnosis was based on intraoperative cholangiography and/or histological confirmation from a liver biopsy obtained during the Kasai procedure.
- Presumed biliary atresia (n = 88): Diagnosis was based on persistent cholestasis along with characteristic ultrasound findings (absence or hypoplasia of the gallbladder, presence of the "triangular cord" sign), high liver stiffness on transient elastography (>20 kPa), absence of intestinal radiotracer excretion on scintigraphy (if performed), and the exclusion of other causes of cholestasis.
- Cholestasis of other etiology (n = 26): Diagnoses in this group were made based on clinical, laboratory, and imaging findings. Specifically:
  - Alagille syndrome was diagnosed when  $\geq 3$  of the following were present: butterfly vertebrae, congenital heart defects, posterior embryotoxon, characteristic facial features, and — when available — JAG1/NOTCH2 gene mutations.
  - PFIC was diagnosed based on clinical features and biochemical markers (including GGT levels); genetic confirmation was performed selectively.
  - Neonatal and CMV hepatitis were diagnosed based on a combination of clinical and laboratory criteria.
  - Choledochal cyst (CDC) was confirmed through ultrasound, MRI, or MSCT data (in cases of intra- and extrahepatic bile duct dilatation).

### **Data Analysis**

Data analysis was performed using IBM SPSS Statistics, version 23. Quantitative variables with normal distribution were presented as mean and standard deviation ( $M \pm SD$ ). Comparisons between patients with presumed BA and those with cholestasis of other etiologies were carried out using the independent Student's t-test. A p-value of <0.05 was considered statistically significant. Correlations between quantitative variables were assessed using Pearson's correlation coefficient.

## **2. RESULTS**

A total of 157 neonates and infants with neonatal cholestasis were included in the study, examined during the period from 2013 to 2023. Based on the results of comprehensive diagnostics, BA (biliary atresia) was either confirmed or considered as presumed in 131 patients (83.4%; Group I), of whom 43 children had the diagnosis verified intraoperatively. In 26 patients (16.6%; Group II), other causes of cholestasis were established. The mean age at the time of BA diagnosis was  $84.3 \pm 3.0$  days; only 15% of children underwent evaluation before reaching the age of two months. In patients with cholestasis of other etiologies, the age at the time of admission was significantly higher ( $p = 0.01$ ). The gender composition of the groups did not differ statistically.

The etiology of cholestasis in Group II (n = 26) was distributed as follows:

- Alagille syndrome was diagnosed in 12 cases,
- Cystic transformation of the choledochus — in 4 patients,
- Neonatal hepatitis (including CMV infection) — also in 4 cases,
- Progressive familial intrahepatic cholestasis (PFIC) — in 3,
- Congenital hepatic fibrosis — in 2,
- Beckwith–Wiedemann syndrome — in 1 case.

**Alagille syndrome** manifested with a typical clinical picture of neonatal cholestasis combined with vertebral body anomalies of the "butterfly" type (based on chest X-ray data), heart defects (tetralogy of Fallot, pulmonary artery stenosis), posterior

embryotoxon (ophthalmologically), and characteristic facial features. Genetic confirmation of the diagnosis was performed selectively.

In four patients, the cause of cholestasis was choledochal cyst (CDC). It was characterized by episodic acholic stools, transient elevation of enzyme levels, and clinical improvement following the use of ursodeoxycholic acid. Differentiation from the cystic form of BA was carried out using ultrasound and MR cholangiography:

in CDC, large cysts of the extra- and intrahepatic bile ducts were visualized, whereas in cystic BA, a small isolated lesion was visualized in the hepatic hilum.

The use of color Doppler mapping (CDM) allowed exclusion of the vascular nature of the formation (Figure 1).



**Fig. 1. Echogram of a 65-day-old infant with the cystic form of biliary atresia (BA). A cystic lesion measuring 0.5 × 0.6 cm is visualized (yellow arrow) in the projection of the right branch of the portal vein.**

In two patients with congenital hepatic fibrosis, hepatomegaly, markedly increased liver stiffness on elastography (>20 kPa), visualization of an anatomically normal gallbladder, and signs of portal hypertension were observed.

In one of these cases, hepatobiliary scintigraphy produced a false-positive result — absence of intestinal excretion of the radiopharmaceutical.

In four children, cholestasis was caused by congenital CMV hepatitis. The disease was characterized by marked cytolysis, moderate cholestasis, normal GGT activity, and normal size and shape of the gallbladder. The diagnosis was confirmed using ELISA (presence of anti-CMV IgM) and PCR.

Distinguishing features included the absence of the "triangular cord" sign on ultrasound, normal GGT levels, and laboratory signs of a viral infection.

Progressive familial intrahepatic cholestasis (PFIC) was diagnosed in three patients. All children had steatorrhea, no signs of obstruction on imaging, and a normal gallbladder structure. GGT levels were normal in two patients and elevated in one. Genetic confirmation of the diagnosis was not obtained in all cases.

Ultrasound examination proved to be an important screening tool. In patients with BA, absence of the gallbladder was noted in 34.5% of cases, marked hypoplasia in 31%; in 35.8% of cases, the gallbladder was visualized with normal shape and size (Fig. 2).



**Fig. 2. Echogram of a 55-day-old infant with biliary atresia (BA). A hypoplastic gallbladder is visualized, without a clearly defined mucosal contour.**

The “triangular cord” sign (an echogenic thickening in the hepatic hilum) was identified in 38.3% of patients with biliary atresia (BA), with an average thickness of  $3.7 \pm 0.18$  mm.

Transient elastography (TE) revealed that liver stiffness in patients with BA was significantly higher than in children with cholestasis of other etiologies ( $21.05 \pm 1.9$  kPa vs.  $17.22 \pm 3.65$  kPa;  $p < 0.05$ ).

A weak but statistically significant correlation was observed between liver stiffness and age ( $r = 0.332$ ;  $p = 0.01$ ), as well as a moderate positive correlation with GGT levels ( $r = 0.36$ ;  $p = 0.007$ ).

No significant correlations were found with bilirubin, transaminases, or platelet counts.

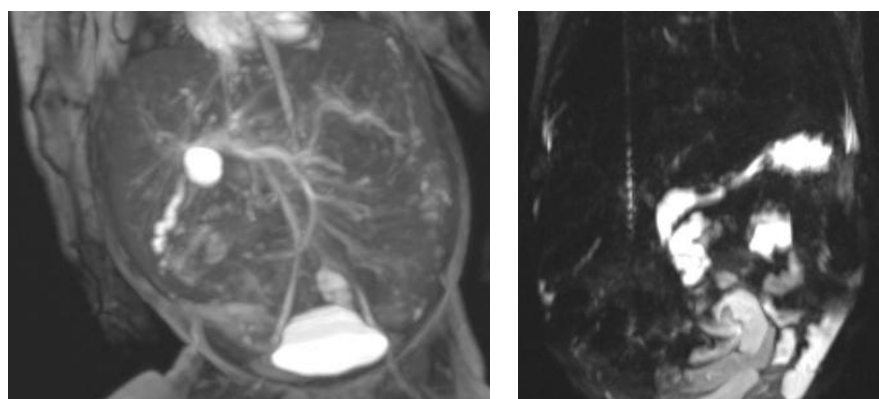
Hepatobiliary scintigraphy was performed in 6 patients with unclear clinical presentations. In all cases, absence of intestinal excretion of the radiopharmaceutical was observed, which was interpreted as a sign of biliary obstruction.

However, the diagnosis of BA was confirmed in only 4 of them (67%), while the other 2 were diagnosed with Alagille syndrome and congenital hepatic fibrosis, respectively.

This supports the high sensitivity but limited specificity of scintigraphy.

MR cholangiography was performed in 12 children and demonstrated high diagnostic value.

- In 4 patients, choledochal cyst (CDC) was identified.
- In 4 others, signs of the cystic form of BA were found.
- In 3 patients, there was a complete absence of bile ducts, consistent with the classic form of BA (Fig. 3).

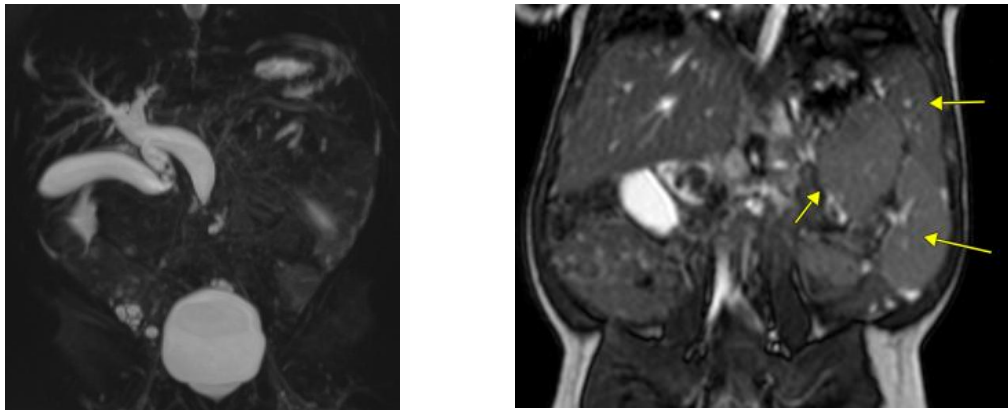


**Fig. 3. MR cholangiography. A – Direct projection, non-contrast cholangiography (MIP reconstruction): a cystic lesion and a deformed gallbladder are visualized in the hepatic hilum. B – MR cholangiogram, direct projection (MIP reconstruction): the gallbladder, as well as the extrahepatic and intrahepatic bile ducts, are not visualized.**

In one case, other diagnostically significant findings were obtained. MR cholangiography proved especially useful for



differentiating choledochal cyst (CDC) from biliary atresia (BA), as well as for excluding anatomical anomalies (Fig. 3).



**Fig. 3. A – MRI with non-contrast cholangiography in a child with cystic transformation of the choledochus, showing fusiform dilation of the choledochus with heterogeneous content in the projection of the cystic duct. B – MRI, T2-weighted image; three spleens (polysplenia) are indicated by arrows.**

### 3. DISCUSSION

The results of this study confirm that biliary atresia (BA) remains the leading cause of neonatal cholestasis, consistent with data from global literature indicating its key role in the structure of obstructive jaundice in newborns [7]. One of the strengths of this study is the use of a comprehensive non-invasive diagnostic approach, including ultrasound, transient elastography (TE), and, when necessary, scintigraphy and MR cholangiography.

Ultrasound proved to be a useful screening method, although its sensitivity was limited: the “triangular cord” sign was identified in less than 40% of BA patients, and a normally shaped gallbladder was visualized in more than one-third of children with confirmed pathology. These findings support previous reports on the dependence of ultrasound accuracy on operator expertise and patient age [16].

The use of TE improved diagnostic accuracy, especially in children over 90 days of age, in whom a significantly increased liver stiffness was observed. However, the interpretation of these data requires caution: liver stiffness values may be influenced by the degree of inflammation, bilirubin levels, and cytolysis severity. Despite a moderate correlation with GGT levels, TE did not show strong associations with other biochemical markers, limiting its standalone prognostic value.

MR cholangiography demonstrated high diagnostic utility in complex differential cases—particularly in distinguishing cystic BA from choledochal cysts. It also proved valuable in identifying associated anomalies (e.g., polysplenia). However, high cost, limited availability, and the need for sedation in neonates hinder its widespread use, especially in resource-limited settings.

Scintigraphy showed high sensitivity, but, as reported previously, its specificity is limited: in our study, two out of six patients with absent intestinal isotope excretion did not have BA. This underscores the need to use scintigraphy strictly based on indications and only in combination with other imaging modalities.

Notably, not all patients with presumed BA underwent surgical confirmation. This is explained, firstly, by delayed presentation—most patients were admitted after 90 days of age, at which point, in our clinical judgment, Kasai surgery was no longer considered rational [22]. Secondly, in some cases, parents, informed about the prognosis, declined surgery, opting instead for delayed primary liver transplantation. These factors should be taken into account when interpreting the results, as some cases lack morphological verification.

Also noteworthy is the relatively high proportion of Alagille syndrome among non-BA cholestasis cases (46.1%), likely reflecting improvements in regional diagnostics, including X-ray, echocardiography, and ophthalmologic screening. Genetic confirmation was performed selectively, which is another limitation of the study. Similarly, in cases of progressive familial intrahepatic cholestasis (PFIC), diagnosis was primarily based on clinical and laboratory findings, as molecular genetic testing was not available in all cases.

General limitations of the study include its retrospective design, inconsistency in diagnostic workups (not all methods were available for every patient), and the relatively small number of non-BA cases, which complicates statistical evaluation of rare cholestatic conditions.

Overall, this study highlights the relevance of a comprehensive diagnostic approach to BA and demonstrates that combining

ultrasound with TE can serve as an effective basis for a non-invasive diagnostic algorithm. A promising direction would be the development and validation of standardized diagnostic protocols for low-resource settings, as well as the implementation of early stool color screening programs, which have proven effective in other countries.

#### 4. CONCLUSION

Biliary atresia remains the leading cause of neonatal cholestasis in Uzbekistan. The combination of ultrasound and transient elastography demonstrates high diagnostic value, especially in children over 90 days of age. However, the limited sensitivity of ultrasound and the variability of TE data necessitate a comprehensive approach using additional imaging modalities. The absence of surgical verification in some patients was due to delayed presentation and parental refusal of surgery in favor of delayed liver transplantation. The findings highlight the importance of early detection of BA, the standardization of diagnostic algorithms, and the improvement of access to specialized care in resource-limited settings.

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