The Egyptian Journal of Hospital Medicine (July 2024) Vol. 96, Page 2950-2958

Role of Ultrasound and Color Doppler in Diagnosis of Biliary Atresia

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ABSTRACT

Background: Biliary atresia is a congenital biliary condition that is defined by an absence or severe deficit of the extrahepatic biliary tree. **Objective:** To detect the role of color Doppler and gray scale ultrasonography as possible tools in evaluation of cases of neonatal jaundice for biliary atresia in comparison to intraoperative cholangiography as a golden test in the diagnosis of biliary atresia.

Patients and Methods: This is a prospective study that included assessment of 100 cases, over a period of two years from the outpatient clinic and the inpatient department of Pediatric Hepatology, gastroenterology and nutrition Department, National Liver Institute, Menoufia University, diagnosed by neonatal cholestaisis.

Results: US score and total biliary atresia score were higher in biliary atresia cases (7.40± 3.69 and 21.73± 3.79, respectively) than non-biliary atresia cases diagnosed by intraoperative cholangiography.

Conclusion: We found that US serves as an important non-invasive imaging modality in diagnosis of BA. In addition to US imaging features as gall bladder abnormalities (small, rudimentary or absent gall bladder), positive TC sign and associated congenital anomalies, color Doppler play an important role in diagnosis of BA as it was found that enlargement of the hepatic artery diameter can be highly suggestive of biliary atresia.

Keywords: Ultrasound, Color Doppler, Biliary atresia, Cholangiogram.

INTRODUCTION

One of the most frequent causes of newborn cholestasis is biliary atresia, which accounts for over half of infants who require liver transplantation and frequently results in cirrhosis that develops right away and results in mortality [(1,2)]. It is estimated that 1 in 10,000–15,000 newborns are affected, with a known male predisposition. In the past fifteen years, there has been a notable rise in the occurrence [(3,4)].

Early detection of biliary atresia should be done as soon as feasible to avoid deadly complications such as malnutrition, liver failure, cirrhosis and its related complications, and it must be distinguished from other non-obstructive causes of neonatal cholestasis to avoid unnecessary surgery [(5,6)]. Diagnosis of biliary atresia depends on clinical picture of obstructive jaundice as color of the skin, the urine and the stool, laboratory by serum total, direct bilirubin and GGT, histopathology to detect ductular proliferation, and imaging abdominal X-ray, ultrasound, MRCP, HIDA scan (cholescintigraphy) and finally confirm diagnosis by intraoperative cholangiography [(7,8)].

Many signs by ultrasound are highly suggestive of biliary atresia as TC sign, subcapsular flow, the absence of the gall bladder, its contractility and its length, hepatic artery diameter and the ratio between it and the portal vein diameter. Ultrasound is the preferred imaging modality for diagnosing biliary atresia due to its low cost, available, rapid, no need to stabilize the child long time as MRCP and no need to use radioactive substance as with HIDA scan [(8,9)].

The aim of this study was to detect the role of color Doppler and gray scale ultrasonography as possible tools in evaluation of cases of neonatal jaundice for biliary atresia in comparison to intraoperative cholangiography as a golden test in the diagnosis of biliary atresia.

PATIENTS AND METHODS

This is a prospective cross-sectional study that included assessment of 100 cases, over a period of two years from the outpatient clinic and the inpatient department of Pediatric Hepatology Department, National Liver Institute, Menoufia University, diagnosed by neonatal cholestaisis. Age of the patients was between 1-100 days. Males and females were included during the period from October 2021 to October 2023.

Exclusion criteria were age above 100 days and refusal of patients' parents to be included in the study.

Study Method:

The included patients underwent:

1.Full history taking.

2.Full clinical examination.

3.Laboratory blood samples examination: (Liver function tests and complete blood picture …etc.).

4.Ultrasonography and color Doppler: examination of the abdomen

A) US examination was the most important and non-invasive imaging procedure during evaluation of cases of BA. All infants had extensive US tests performed by a single operator utilizing a US system (Toshiba, Xario, USA) equipped with a 4.5-MHz curvilinear transducer and a 12-MHz linear array transducer.

B) The following features were recorded:

1- Liver size and echo-texture: The transducer selected was a curved transducer a 4.5 MHz however; linear transducer was used also for more detailed imaging of the hepatic parenchyma. The patient was

Received: 03/03/2024
Accepted: 05/05/2024

generally supine in position, the liver size was measured using longitudinal scan in the mid clavicular line where the right hepatic lobe should not extend more than 1 cm below the costal margin in young infants.

2-Gallbladder length and morphology: Using curvilinear transducer 12-MHz linear-array transducer placed over the right upper quadrant of the upper abdomen to assess the shape and irregularity of the wall of the gall bladder as well as its length using more oblique scans to measure the gall bladder length at its large dimension. Also the gall bladder was evaluated for the presence of sludge, stones or pericholecystic collection.

3-The triangular cord sign: It was assessed by sing 12-MHz linear-array transducer. This sign was imaged by scanning in both transverse oblique plane or longitudinal plane and identified as focal area of increased echogenicity anterior to bifurcation of portal vein that appeared as a triangular cone shaped echogenicity anterior to the right portal vein in some cases while in the other cases it was cord like. In all cases the maximum thickness of the TC sign was measured. A thickness of more than 4 mm was regarded as a positive TC sign because it indicates the thickness of any normal structures that may have existed along the anterior aspect of the portal vein, such as the common hepatic duct, the right hepatic artery's anterior wall, and the right portal vein's anterior wall.

4-Spleen size and morphology: The transducer was placed in the left upper abdomen in an oblique manner for measurement of the spleen size, also the spleen was assessed for presence of polysplenia or splenic malformations as these findings are common association in cases of biliary atresia.

5-Hepatic artery and portal vein diameter and color Doppler examination of their flow: Both hepatic artery and portal vein diameters were measured near the porta hepatis using linear transducer in transverse oblique plane. Also color Doppler ultrasound was used to determine the presence and the direction of blood flow.

6-The presence of hepatic subcapsular flow: The hepatic subcapsular flow was assessed by placing the linear transducer in transverse position over the right hypochondrium and the image depth reduced to allow good focus over the subcapsular region then the color box was placed over the subcapsular region. When vascular structures on color Doppler US pictures continued to the liver capsular surface, we regarded this as evidence of the presence of the hepatic subcapsular flow.

7-The presence of any associated congenital anomalies: Following the completion of US, all patients underwent an evaluation to determine whether any congenital malformations were connected with BA. Examples of circulatory anomalies include polysplenia, situs inversus, an interrupted inferior vena cava, and others.

5-Percutaneous US guided needle biopsy:

A) Percutaneous needle biopsy was conducted on all 100 patients who were suggested to be diseased with BA. The specimen was taken under ultrasound guidance with complete aseptic conditions and prebiopsy sedation. This fine needle biopsy was taken with gun needle or true cut needle and their caliber 1.0, 1.2 and 1.4 mm with beveled tip at 45 degree and flat ground.

B) The needle was introduced under ultrasound guidance and its tip was followed till the region of interest keeping away from vascular structure and gall bladder. The specimen was installed in saline and sent to the Pathology Department as soon as possible.

C) Post biopsy US examination was done to evaluate any possible complication, where the histological appearance of the liver biopsy is characterized by bile duct clogging, ductal growth, portal edema, small cell infiltration, and variable giant cell development.

6-Intraoperative Cholangiography:

A) Intraoperative cholangiography (IOC) was the most conclusive technique for intraoperative assessment of the biliary atresia. It was done for all cases suspected to be biliary atresia in the operating room for proving the diagnosis and deciding the exact mode or subtype of the operation that suited every case.

B) After dissection in Calot's triangle, the surgeon introduced a catheter into the biliary tree, then irrigated with saline to ensure no air bubbles. All surgical instruments were removed from field of exposure (except fixing clamps).

C) Fractionated dose of hypaque 25% or urografin 30% was injected at 3, 5, 8 ml even 10 ml till filling the field and serial (subtracted) X-ray fluoroscopy images were obtained. The use of dynamic fluoroscopy had improved the speed with which intraoperative cholangiography could be performed and yielded a series of high-resolution images that more accurately identified the different surgical types of biliary atresia and hence the surgeon could decide the subtype of the operation that suited every case.

Ethical approval:

The Institution Review Board of the Faculty of Medicine at Menoufia University approved the study. All patients' parents provided informed permission following a thorough explanation of the advantages and hazards. All data were kept private, and each patient file was assigned a code number that contained all of the research conducted. The Helsinki Declaration was observed throughout the study's duration.

Statistical analysis

Using Windows SPSS version 22, the collected data were coded, processed, and examined. A normal distribution of the data was checked using the Shapiro-Wilk test. The frequency and relative percentages of the qualitative data were presented and compared by the X^2-test. Mean ± standard deviation (SD) and range were used to report quantitative data. Two independent groups of parametric data that were normally distributed were compared using an independent samples t-test. ROC curve analysis evaluated a test's diagnostic performance, or its ability to distinguish between sick and non-diseased cases. P-values less than or equal to 0.05 were considered significant.

RESULTS

1. Socio-demographic data of the participants (N=100)

This is a prospective study that was conducted over 100 patients at national liver institute. Mean age of the patients was 66.36±13.0. 55 patients were females. All the patients showed clay stool (Table 1).

Table (1): Socio-demographic data of the participants (N=100):

	Distribution (N=100)		
Age(days):	Mean± SD	66.36±13.0	
	Range	38-92	
		N	**%**
Sex:	Male	45	45.0
	Female	55	55.0
Clinical data (clay stool):	Yes	100	100.0

2. Lab investigations of the participants (N=100)

CBC of the patients showed that the mean hemoglobin value of the patients was 9.3±1.4 (range between 6.7-12.3). Mean platelet count was 296.6±140 (range between 62.0-659.0). Mean WBCs count was 10.9±4.5 (range between 3.2-28.7). Mean PC count was 77.4±16.3 (range between 45.0-130.0).

Liver function test showed that mean total bilirubin level was 9.7±3.7 (range between 4.5-23.3), mean direct bilirubin level was 7.3±4.5 (range between 2.2-38.0), mean SGOT level was 160.2±104.8 (range between 33.0-480.0), mean SGPT level was 123.7±108.3 (range between 28-504), and mean GGT level was 585.2±448.5 (range between 51-2115).

Kidney function test showed that mean creatinine level was 0.4±0.2 (range between 0.1-1.3).

3. Ultrasound results:

US results showed that mean liver size (cm) was 9.1±1.7, mean spleen size (cm) was 6.5±1.5, mean hepatic artery diameter (HAD) (mm) was 1.9±0.7, mean portal vein diameter (PVD) (mm) was 4.6±0.7, and mean HAD/PVD (mm) was 0.4±0.1. Mean GB length before feeding (mm) was 22.3±10.1 and after feeding (mm) was 14.3±7.7. Gall bladder contractility was contractile in 34 patients (34%) and negative in 66 patients (66%). IHBRDS was not found in 86 patients (86%). TC sign was positive in 8 patients (8%). Subcapsular flow was positive in 17 patients (17.0%). Ascites was rim in 4 patients (4%), minimal in 13 patients (13%), and mild in 3 patients (3%) (Table 2).

Table (2): US results:

Distribution (N=100)		
	Mean±SD	**Range**
Liver size(cm)	9.1±1.7	4.5-12.0
Spleen size (cm)	6.5±1.5	4.2-11.7
HAD (mm)	1.9±0.7	1.0-4.0
PVD (mm)	4.6±0.7	3.0-6.6
HAD/PVD (mm)	0.4±0.1	0.16-0.80
GB before feeding(mm)	22.3±10.1	6.0-40.0
GB after feeding (mm)	14.3±7.7	4.0-36.0
	N	**%**
TC sign:		
Positive	8	8.0
Negative	92	92.0
Subcapsular flow:		
Positive	17	17.0
Negative	83	83.0
Gallbladder contractility:		
Contractile	34	34.0
Negative	66	66.0
IHBRDS:		
Left	4	4.0
Minimal	10	10.0
No	86	86.0
Ascites:		
Rim	4	4.0
Minimal	13	13.0
Mild	3	3.0
No	80	80.0

4. According to El-Guindi biliary atresia score (Table 3):

100 patients had clay stool (score=2.907). By US, TC sign was positive in 8 patients (score=2.418). Contractile gall bladder was negative (score=2.773) in 66 patients. Gall bladder length before feeding (mm) was <20 (score=2.576) in 62 patients. HAD (mm) was >2.05 (score=2.037) in 20 patients. HAD/PVD (mm) was >0.445 (score=1.705) in 42 patients. Subcapsular flow was positive in 17 patients (score=6.735).

Laboratory results were platelet (*103) score was >349 (score=1.417) in 32 patients. GGT score was >286 (score=2.576) in 75 patients. By histopathology, we found that ductular proliferation was positive in 74 patients (score=5.239), bile plugs were positive in 97 patients (score=3.914), and multinucleated giant hepatocytes were negative in 47 patients (score=2.883) (Table 4).

Table (3): El-Guindi Biliary atresia score [10]:

	Variable	Score if Yes	Score if No
Clinical			
1	Clay stool	2.907	0
Ultrasonographic			
2	Triangular cord sign	2.418	0
3	Contractile gallbladder	0	2.773
4	Gallbladder length ≥20.5 mm	0	2.576
5	HAD ≥2.05 mm	2.037	0
6	HAD/PVD ≥0.445 mm	1.705	0
7	Hepatic subcapsular flow	6.735	0
Laboratory			
8	γGT ≥286 U/L	2.576	0
9	Platelets ≥349 x 10^3/µl	1.417	0
Histopathology			
10	Ductular proliferation	5.239	0
11	Bile plugs	3.914	0
12	Multi-nucleated giant hepatocytes	0	2.883

Maximum score = 37.18
>23.927 means BA
<23.927 means non-BA

Table (4): Diagnosis of biliary atresia by different parameters:

		N	%
	Clinical		
Clay stool:	Yes score=2.907)	100	100.0
	US		
TC sign:	Positive (score=2.418)	8	8.0
	Negative(score=0)	92	92.0
Contractile Gallbladder:	Positive(score=0)	34	34.0
	Negative(score=2.773)	66	66.0
Gallbladder before(mm):	>20 (score=0)	38	38.0
	<20(score=2.576)	62	62.0
HAD (mm):	>2.05 (score=2.037)	20	20.0
	<2.05 (score=0)	80	80.0
HAD/PVD (mm):	>0.445(score=1.705)	42	42.0
	<0.445 (score=0)	58	58.0
Subcapsular flow:	Positive (score=6.735)	17	17.0
	Negative(score=0)	83	83.0
	Laboratory		
Platelet (*10^3) score:	>349(score=1.417)	32	32
	<349(score=0)	68	68
GGT score:	>286 (score=2.576)	75	75
	<286 (score=0)	25	25
	Histopathology		
Ductular proliferation:	Yes (score=5.239)	74	74.0
	No(score=0)	26	26.0
Bile plugs:	Yes (score=3.914)	97	97.0
	No(score=0)	3	03.0
Multinucleated giant hepatocytes:	Yes (score=0)	53	53.0
	No(score=2.883)	47	47.0

5. Total Biliary atresia score and its components:
The mean clinical score was 2.91 ±0. The mean laboratory score was 2.39 ±1.30. The mean US score was 5.89 ±4.23. The mean histopathology score was 7.52±2.76. The mean total biliary atresia score was 18.70 ±6.18 (Table 5).

6. Final Diagnosis and Evaluation:
The biliary atresia by cholangiogram intraoperative was positive in 74 patients (74%) and negative in 26 patients (26%). Type of biliary atresia was Type IIb in 8 patients (10.8%) and Type III in 66 patients (89.2%). Kasai operation was done on 63 patients (63%) and not done on 37 patients (37%).

7. Comparison between biliary atresia and non-biliary atresia cases regarding different diagnostic parameters:
Regarding comparison between biliary atresia and non-biliary atresia cases, we found that there was no significant difference between both groups regarding TC sign, HAD, HAD/PVD, platelets score and GGT score (P. value > 0.05). There was significant difference between both groups regarding subcapsular flow (P. value 0.005). There was highly significant difference between both groups regarding contractile gallbladder, gallbladder before feeding, ductular proliferation, bile plugs, and multinucleated giant hepatocytes (P. value < 0.001).

In biliary atresia group (46 patients), mean GB length before feeding was 19.3 ± 10.7 with range between 6.0 – 40.0. In non-biliary atresia group (26 patients), mean GB length before feeding was 27.4 ± 6.3 with range between 15.0 –38.0.

8. Comparison between biliary atresia and non-biliary atresia cases regarding total biliary atresia score and its components:
There was highly statistically significant difference between biliary atresia and non-biliary atresia cases regarding US score and total biliary atresia score (P<0.001). US score and total biliary atresia score were higher in biliary atresia cases (7.40± 3.69 and 21.73± 3.79, respectively) than non-biliary atresia cases diagnosed by intraoperative cholangiography (Table 5).

Table (5): Comparison between biliary atresia cases and non-biliary atresia cases regarding biliary atresia score and its components:

	Biliary atresia (N=74)	No biliary atresia (N= 6)	Mann-Whitney test	P-value
	Mean ± SD Range	Mean ± SD Range		
Clinical score	2.91±0 2.91	2.91±0 2.91	-	-
Laboratory score	2.27±1.31 0–3.99	2.72±1.24 0–3.99	1.66	0.096
US score	7.40±3.69 1.71–15.83	1.59±2.26 0–6.32	6.24	**<0.001 (HS)**
Histopathology score	9.15±0 9.15	2.88±0 2.88	-	-
Total Biliary atresia score	21.73±3.79 13.77-30.46	10.10±2.31 5.79–14.68	**T- test** 18.42	**<0.001(HS)**

Cut-off point was > 3.258 with sensitivity of 89.2 % and specificity of 80.8% (Table 6 and Figure 1).

Table (6): ROC curve for US score as a predictor for biliary atresia diagnosis

Cut-off point	AUC	Accuracy	Significance	Sensitivity	Specificity	95%CI
>3.258	**0.908**	**%90.8**	**<0.001**	**89.2 %**	**80.8 %**	**0.84-0.98**

AUC: area under curve**; CI:** confidence interval (lower-upper).

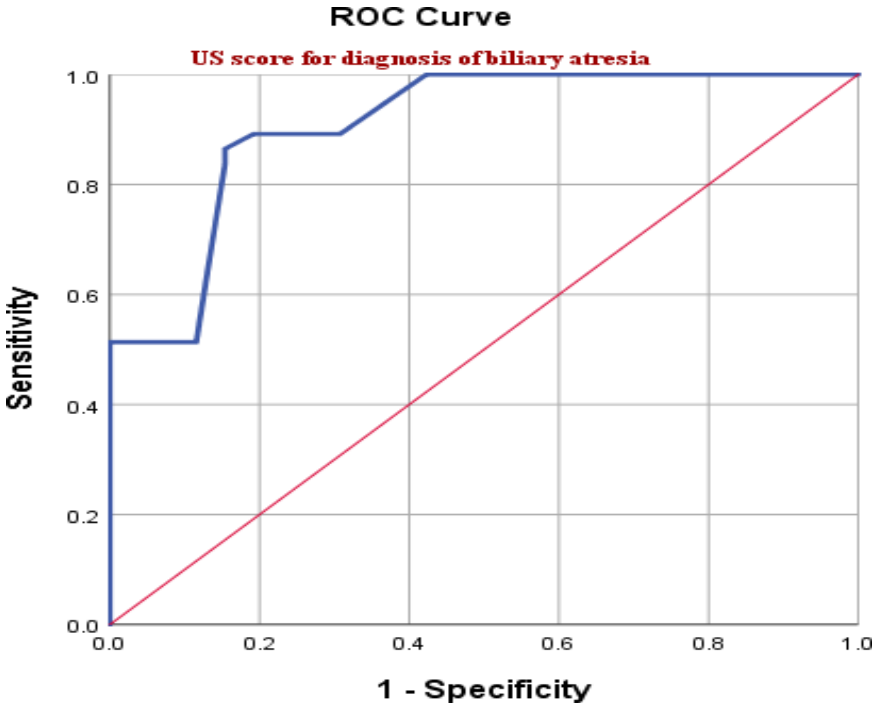

Figure (1): Showing ROC curve for US score as a predictor for biliary atresia diagnosis

CASES

CASE 1

A female infant aged 54 days old, jaundiced at the 6th day of birth with a progressive course. US examination revealed: left segmental IHBRDs, contracted gall bladder, porta hepatis cystic lesion 6.4x5.7 mm, and positive subcapsular flow. MRCP showed non-visualized gall bladder, porta hepatis cystic structure measuring 5 mm in diameter and non-visualized extrahepatic biliary system. This case was biopsied and its specimen was compatible with biliary atresia. Intraoperative cholangiography was done as an important confirmatory step during the operation, showing Kasai classification type IIa (cystic biliary atresia). (Figure 2)

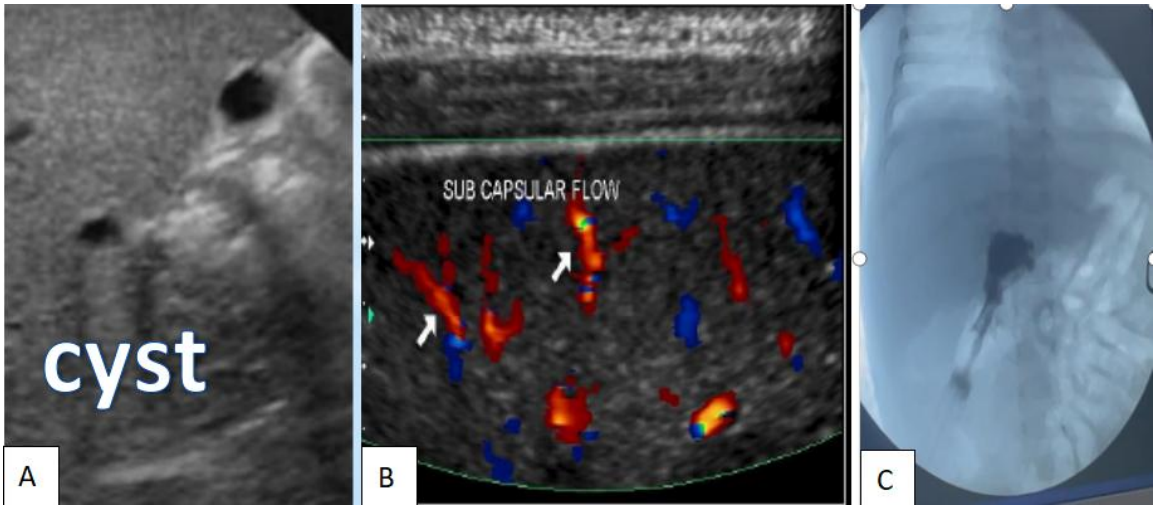

Figure (2): A female infant aged 54 days old, jaundiced at the 6th day of birth with a progressive course. (A) US showed contracted gall bladder and porta hepatis cystic lesion 6.4x5.7mm. (B) Positive subcapsular flow. (C) Intraoperative cholangiogram showing Kasai classification type IIa (cystic biliary atresia).

CASE 2

A male infant aged 53 days old, presented with jaundice at the 3rd day of birth with a progressive course and clay stool. US and color Doppler revealed absence of gall bladder, positive TC sign measuring about 4.3 mm thickness, an enlarged hepatic artery measuring about 2.9 mm in diameter, associated congenital anomaly (polysplenia), and prominent hepatic subcapsular flow is seen on color Doppler US. On biopsy, the specimen gave positive result for biliary atresia and was confirmed by intraoperative cholangiography (Figure 3).

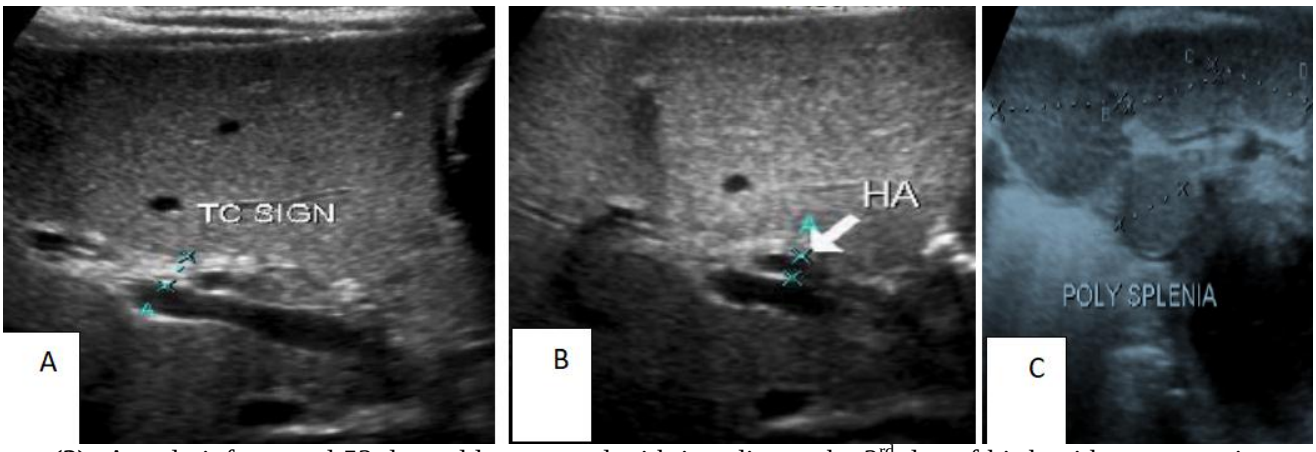

Figure (3): A male infant aged 53 days old, presented with jaundice at the 3rd day of birth with a progressive course and clay stool. (A) US revealed absence of gall bladder, positive TC sign measuring about 4.3 mm thickness. (B) An enlarged hepatic artery measuring about 2.9 mm in diameter and associated congenital anomaly (polysplenia).

DISCUSSION

Since most preterm and over half of full-term infants suffer from neonatal cholestasis, it is important to watch for signs of hyperbilirubinemia in newborns to avoid acute bilirubin encephalopathy [11,12]. The yellowish staining of a newborn baby's skin and other tissues is known as neonatal jaundice. In infants, a bilirubin level exceeding 5 mg/dL results in clinical jaundice, but in adults, a level of 2 mg/dL might appear icteric [12,13].

In our study, mean age of the patients was 66.36±13.0 (range between 38-92 days). 45 patients were males and 55 patients were females. 100 patients showed clay stool.

Our results agreed with the study by **Humphrey and Stringer** [14], as the mean age of patients at US assessment was 52.4 ±27 days, with (P >0.5), while **Zhang *et al.*** [13] found that diagnosis was confirmed in infants and children aged 2-20 months. The incidence of BA was shown to be higher in females (1.62 per 10,000) than in boys (1.30 per 10,000) by **Davenport *et al.*** [15], but the difference wasn't statistically significant.

We agreed with **Shi *et al.*** [16], who mentioned that females were affected slightly more often than males, **Davenport *et al.*** [15] confirmed that there was usually a female predominance in BA cases. While **Schreiber *et al.*** [17], found that sex distribution in BA was equal.

We agreed with **Lee *et al.*** [18], results which reported that based on consensus reading, hepatic subcapsular flow had sensitivity, specificity, positive and negative predictive values of 100%, 86%, 85%, and 100%, respectively. Subcapsular telangiectatic vessels were present in all BA patients with hepatic subcapsular flow on color Doppler US pictures at the time of the Kasai surgery.

In accordance with our study, **Humphrey and Stringer**[14] reported that in comparison to neonates without BA (25.9 mm ± 6.9), the mean gallbladder length was considerably shorter in those with BA (18.8 mm ± 7.9).

However, these are rather general results, thus a diagnosis of BA cannot be established based only on them. These findings concur with those of **Humphrey and Stringer**[14], who found that three of the four newborns diagnosed with BA splenic malformation syndrome also had an interrupted retrohepatic vena cava at US and polysplenia. At laparotomy, the fourth patient's segmentation spleen and congenital portocaval fistula were discovered, along with the absence of a portal vein.

In our study, two cases only had polysplenia and no other congenital anomalies were detected.

Liver biopsies might be carried out per US guidelines or in the operating room. Pathological investigation reveals a lack of bile ducts, bile plugs, bile duct growth due to biliary obstruction, and potential abnormalities of the ductal plate [17,18].

In our study, histopathological examination showed that ductular proliferation was positive in 74 % and negative in 26%, bile plugs were positive in 97% and negative in 3%, and multinucleated giant hepatocytes were positive in 53% and negative in 47%.

When it comes to diagnosing BA, intraoperative cholangiography is considered the gold standard and the conclusive investigative test. Cholangiograms, however, have been shown in multiple trials to be unreliable in differentiating between patients with and without BA. This resulted in the incorrect diagnosis of non-BA patients as BA patients, which could be a significant factor in the disparate postoperative results of Kasai that various hospitals reported [18,19].

In our study, the biliary atresia by cholangiogram intraoperative was positive in 74 patients (74%) and negative in 26 patients (26%). Type of biliary atresia was Type IIb in 8 patients (10.8%) and Type III in 66 patients (89.2%). Kasai operation was done on 63 patients (63%) and not done on 37 patients (37%).

There was highly statistically significant difference between biliary atresia and non-biliary atresia cases regarding US score and total biliary atresia score (P<0.001). US score and total biliary atresia score were higher in biliary atresia cases (7.40± 3.69 and 21.73± 3.79, respectively) than non-biliary atresia cases diagnosed by intraoperative cholangiography.

Using a clever statistical technique, **El-Guindi *et al.*** [10] created a new twelve-point diagnostic score based on the most recent information available regarding all previously documented indicators of BA. A group of 75 consecutive patients underwent subsequent validation, which revealed an overall high diagnosis accuracy of 98% (sensitivity 100%, specificity 97.6%).

In the study by **Sciveres *et al.*** [20], neonatal cholestasis resulted in the referral of 64 individuals. Eighteen of them received liver biopsy, two endoscopic retrograde cholangiopancreatography procedures, and eighteen intraoperative cholangiography procedures following initial assessment, which included clinical examination, ultrasonography, and targeted laboratory and/or radiological tests to rule out alternative definitive diagnosis.

After a median delay of 6 days from the initial evaluation (range 1–47 days), 16 babies with the diagnosis of BA had the Kasai surgery at a median age of 62 days (range 34–128 days). After the initial assessment, just three patients had an incorrect diagnosis [20].

After 47 days, one BA patient underwent intraoperative cholangiography, as the initial diagnosis of cholestasis was due to concurrent hypothyroidism. On the other hand, two infants underwent intraoperative cholangiography and were subsequently diagnosed with neonatal sclerosing cholangitis and non-syndromic ductal paucity, respectively [20].

In 27 patients (11 from surgery and 16 from needle biopsy), liver histology was available for examination. They determined the **El-Guindi *et al.*** [10] diagnostic score at the time of the initial evaluation in hindsight for that group of 27 patients.

Mean age at first evaluation of the 27 infants was 60.4 days (range 29–122 days), 16 children had BA and 11 another cause of neonatal cholestasis (3 transient neonatal cholestasis, 1 cystic fibrosis, 1 neonatal sclerosing cholangitis, 1 hypothyroidism, 2 non-syndromic ductopenia, 1 mitochondrial depletion syndrome, 1 Niemann-Pick type C, 1 biliary acids synthesis defect) [10]. Only the gallbladder's contractility could not be assessed using the captured pictures. This item was calculated by comparing the initial examination—conducted following a fast—with additional, haphazardly conducted examinations in the days that followed. For every patient, there was at least one follow-up ultrasound available [10, 20].

Global sensitivity was 31% (5/16), specificity was 90.9% (10/11), and negative and positive predictive values were respectively 0.47 and 0.81. The overall result for diagnostic accuracy was 0.55 [10, 20].

Numerous factors could be at blame for these unsatisfactory outcomes. First, a significant complicating issue was the study's retrospective design. To validate that score, a sizable prospective research with multiple focus points was required. However, they felt that the patient group they employed was very different from the one the authors used to validate the score [21, 22].

In our study, according to this biliary atresia score by **El-Guindi *et al.*** [10]**,** our study found that 100 patient have had clay stool (score=2.907). By US, TC sign was positive in 8 patients (score=2.418) and negative in 92 patients (score=0). Contractile Gall bladder was positive (score=0) in 34 patients and negative (score=2.773) in 66 patients. Gall bladder length before feeding (mm) was > 20 (score=0) in 38 patients and <20 (score=2.576) in 62 patients. HAD (mm) was >2.05 (score=2.037) in 20 patients and <2.05 (score=0) in 80 patients. HAD/PVD (mm) was >0.445 (score=1.705) in 42 patients and <0.445 (score=0) in 58 patients. Subcapsular flow was positive in 17 patients (score=6.735) and negative in 83 patients (score 0).

Laboratory results were platelet (*103) score was >349 (score=1.417) in 32 patients and <349 (score=0) in 68 patients. GGT score was >286 (score=2.576) in 75 patients and <286 (score=0) in 25 patients.

By Histopathology we found that ductular proliferation was positive in 74 patients (score=5.239) and negative in 26 patients (score=0), bile plugs were positive in 74 patients (score=3.914) and negative in 26 patients (score=0), and multinucleated giant hepatocytes were positive in 26 patients (score=0) and negative in 74 patients (score=2.883).

The mean clinical score was 2.91 ±0. The mean laboratory score was 2.39 ±1.30 (range between 0 – 3.99). The mean US score was 5.89 ±4.23 (range between 0–15.83). The mean histopathology score was 7.52±2.76 (range between 2.88-9.15). The mean Total biliary atresia score was 18.70 ±6.18 (range between 5.79–30.46).

So, this scoring system is shown to be an effective reliable method to detect BA in neonates and it will lead to early diagnosis and management of BA.

RECOMMENDATIONS

*Combination between clinical and radiological evaluation is very important for diagnosis of BA.
*US findings with histopathological results and intraoperative cholangiogram can confirm the diagnosis of BA.
*EL-Guindi *et al*. biliary atresia score is a very important score that can be used for diagnosis of BA.

LIMITATIONS

*The study was conducted over a relatively small number of patients. It was confined to a single center (National Liver Institute, Menoufia University Hospitals).
*Patients' parents' refusal to join our study.

CONCLUSION

*We found that US serves as an important non-invasive imaging modality in diagnosis of BA. In addition to US imaging features as gall bladder abnormalities (small, rudimentary or absent gall bladder), positive TC sign and associated congenital anomalies, color Doppler play an important role in diagnosis of BA as it was found that enlargement of the hepatic artery diameter can be highly suggestive of biliary atresia.

*Also, the presence of hepatic subcapsular flow is an essential imaging finding in patients with biliary atresia, which represents the perivascular arterial tuft that surrounds the irregular or blocked peripheral arterial segments that are detected in cases of BA.
*The gold standard for diagnosing biliary atresia is a cholangiogram. Laparotomies or percutaneous methods can be used to do this surgery. The best is to inject contrast agent into the gallbladder whenever feasible. A small atretic gallbladder, the absence of the gallbladder lumen, or the lack of contrast agent in the biliary ducts and small bowel are all signs of biliary atresia.

- **No funding.**
- **No conflict of interest.**

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