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Contribution of Ultrasound in the Diagnosis of Biliary Atresia in Mali

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Abstract **Original Research Article**

Introduction: Biliary atresia is a congenital malformation resulting from an inflammatory, destructive and sclerosing process, affecting the intra and extra hepatic bile ducts. It is the leading surgical cause of neonatal cholestasis and liver transplantation in children, of poorly understood etiology. Clinical diagnosis is suggested by jaundice, discolored stools and dark urine. Ultrasound is essential for diagnosis. The objective is to study the contribution of ultrasound in diagnosis. ***Materials Methods:*** This was a retrospective and prospective bi-center cross-sectional study (CHU-Luxemburg and CHU-Kati) carried out from January 2019 to May 2022, i.e. 41 months. The equipment used was branded devices: EDAN and MINDRAY equipped with three probes with Doppler. ***Results:*** A total of 8 cases out of 2200 pediatric ultrasounds from 0-06 months, representing a frequency of 0.36%. The average age was 45.5 days with extremes of 07-90 days. The age group 08-28 days was more represented (62.5%), the sex ratio was 0.6. All patients were jaundiced with biological cholestasis. The bile ducts were absent on ultrasound in all patients; the gallbladder was present in two patients, with an interruption of the gallbladder lumen in one patient and the presence of a cord sign (37.5%). Management was surgical. ***Conclusion:*** Biliary atresia is a rare condition. Prenatal ultrasound diagnosis is exceptional; in our setting, it is made after birth. Treatment is surgical.

Keywords: Biliary atresia - pediatrics - ultrasound - Kati and Luxembourg University Hospitals.

INTRODUCTION

Biliary atresia (BVA) is a congenital malformation resulting from an inflammatory, destructive, and sclerosing process affecting the intra- and extrahepatic bile ducts. This process leads to fibrosis and obliteration of the bile ducts, resulting in, the development of cholestasis, then liver cirrhosis [1,2]. It is the leading surgical cause of neonatal cholestasis and liver transplants in children [3].

Its incidence is 1/1,800 births in Europe, or approximately 45 cases per year in France [3]. Girls appear to be more often affected than boys, with a female sex ratio of 1.6 [4].

In Mali, a study conducted by Aremu YA *et al.,* [3], in 2020 in the pediatric surgery department of Gabriel Toure University Hospital, found a hospital frequency of 0.25% with a prevalence of 3 cases/year.

The cause of AVB is poorly understood. The pathogenic mechanisms involved could reveal the interaction between different environmental factors. Some authors, such as Strickland in 1982, in an epidemiological study conducted in North Texas, revealed the possibility of a toxic agent being used [5]. Others have reported an ischemic origin with a possibility of the association of AVB with cytomegalovirus, respiratory syncytial virus, Epstein Barr virus, and human papillomavirus [6]. Still others such as Urushihara support an immunological and genetic origin [7].

In approximately 10% of cases, biliary atresia is said to be syndromic, accompanied by other malformations such as polysplenia (or more rarely asplenia), situs inversus, a median liver, intestinal malrotation, a preduodenal portal vein and inferior vena cava agenesis [1]. Most often, in 90% of cases, it is a non-

syndromic atresia, in which the biliary anomaly is isolated.

The clinical diagnosis of BVA is suggested by the classic triad (jaundice that persists after two weeks of life, discoloured stools and dark urine); hepatomegaly [1], and biology which only confirms the diagnosis of hepatic cholestasis, in front of the increase in the serum level of predominantly conjugated bilirubin (greater than 20% of total bilirubin), alkaline phosphatases, cholesterol, as well as the remarkable rise in the level of gamma-glutamyl-transferases (up to 10 times normal). Transaminases are moderately increased (4 to 5 times normal) [6].

Ultrasound provides an important preliminary study in the assessment of this pathology. The authors propose a diagnostic strategy in the evaluation of infantile cholestasis by emphasizing the sonographic cord sign as the first priority of investigations. When the triangular cord sign is visualized, rapid exploratory laparotomy is mandatory without further investigations. In the absence of this sign, other investigations are necessary, including MRI, which, despite its potentially wide application in pediatric patients, affects the biliary system revolution due to the small size of the ducts, the average partial volume, and the blurring of movement related to respiratory and cardiac movements. It is a very reliable non-invasive imaging modality for the diagnosis of biliary atresia.

To our knowledge, this study is the first in Mali, but other clinical and surgical studies have been conducted.

MATERIALS AND METHODS

This was a retrospective and prospective cross-sectional study conducted across two centers (CHU-Luxemburg and CHU-Kati) from January 2019 to May 2022, a period of 41 months. The equipment used was EDAN and MINDRAY, installed in 2016 and 2018 respectively. These devices were equipped with three probes (7.5-13 MHz linear, 3.5 MHz convex, and endocavitary) with color and pulsed Doppler. The examinations were performed by radiologists; each patient underwent at least two ultrasounds to confirm the clinical diagnosis. Patients were fasting for 2 to 4 hours. The duration of the examination depended on the patients' age. The patient was placed in a room with sufficient dim light. Parents actively participated to reduce the children's anxiety and to immobilize the child to improve the quality of the examination. The children were undressed down to the pubic symphysis; the diaper was partially undone to avoid any unforeseen urinary flooding. A bottle of fruit juice or a few mouthfuls of breast milk or sweetened water could be given during the examination. The examination was performed without sedation with a low-frequency 3.5 MHz probe, then with a high-frequency probe (7-13 MHz). It involved all compartments of the abdomen. The parameters studied were: age, sex, pregnancy history, clinical, biological and ultrasound data. In the ultrasound data, the emphasis was placed on the absence of the bile ducts, the absence of the gallbladder or irregularity of the gallbladder wall if present, the cord sign. We also looked for situs inversus, polysplenia, the continuation of the azygos vein with the inferior vena cava, a preduodenal portal trunk, and inversion of the mesenteric vessels (intestinal malrotation). All infants aged 0 to 6 months who had a clinical or clinical-biological syndrome of cholestatic jaundice and in whom ultrasound diagnosed biliary atresia were included.

Ethical aspect: Informed verbal consent was obtained from parents, respecting anonymity, for all patients.

RESULTS

At the end of our study, we recorded 8 cases of AVB, 5 at the Luxembourg Mother and Child University Hospital and 3 at the Kati University Hospital, out of a total of 2,200 pediatric ultrasounds, representing a frequency of 0.36%. The mean age was 45.5 days, with a range of 7-90 days (Table I). They involved 5 girls and 3 boys, representing a sex ratio of 0.6. These were children from well-monitored, full-term pregnancies whose mothers all had a third-trimester ultrasound, revealing no abnormalities. Cholestasis syndrome (jaundice, discolored stools, and dark urine) was present in all our patients. Biologically, cholestasis was found in all our patients, signs of cytolysis and hepatocellular insufficiency in 2 patients (25%) and anemia in 6 patients (75%). On ultrasound (Table II), the intrahepatic bile ducts were not visible in any patient, the gallbladder was visible in 2 patients (25%) with thick and irregular walls, the cord sign was present in 3 patients (37.5%). Peri-gallbladder fibrosis was visible in 2 patients, or 25%, as well as cysts/microcysts. Polysplenia was noted in a single patient who also presented situs inversus, with inversion of the mesenteric vessels (intestinal malrotation). Management was surgical in all cases. The postoperative course was simple in 6 patients; we deplore 2 cases of death due to hepatic cytolysis.

Table I: Distribution of patients according to sociodemographic data:

Sociodemographic Data	Number	Percentage
Age		
Early neonatal 0-7 days	01	12,5 %
Late Neonatal 8-28 days	05	62.5%
Infant 1-3 months	02	25%
Sex		
Early Neonatal 0-7 days	03	37,5%
Late Neonatal 8-28 days	05	62,5%

Table II: Distribution according to ultrasound signs of AVB

Ultrasound signs	Present		Absent	
	Effectif	Pourcentage	Effectif	Pourcentage
Gallbladder	02	25%	06	75%
Wall Irregularity	02	25%	06	75%
Lumen	01	12,5%	06	75%
Perigallbladder Fibrosis	02	25%	06	75%
Intrahepatic Bile Ducts	00	-	08	100%
Cord Sign	03	37,5%	05	62,5
Hilar Fibrosis	03	37,5%	05	62,5%
Cyst and Microcyst	02	25%	06	62,5
Associated signs: polysplenia Situs inversus and ascites	01	12,5%	00	-

Iconography: We present some images for illustrative purposes.

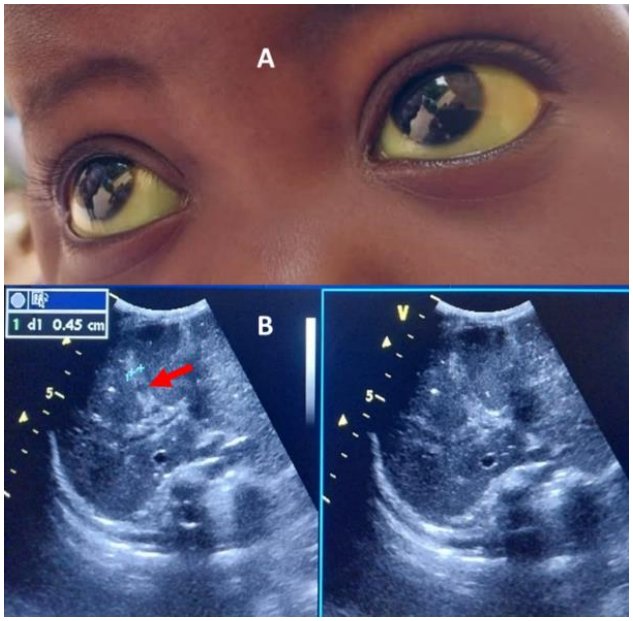

Figure 1: Photograph of a 3-month-old female infant showing frank jaundice (A), Infant liver ultrasound showing a gallbladder with an interrupted lumen (red arrow), (B)

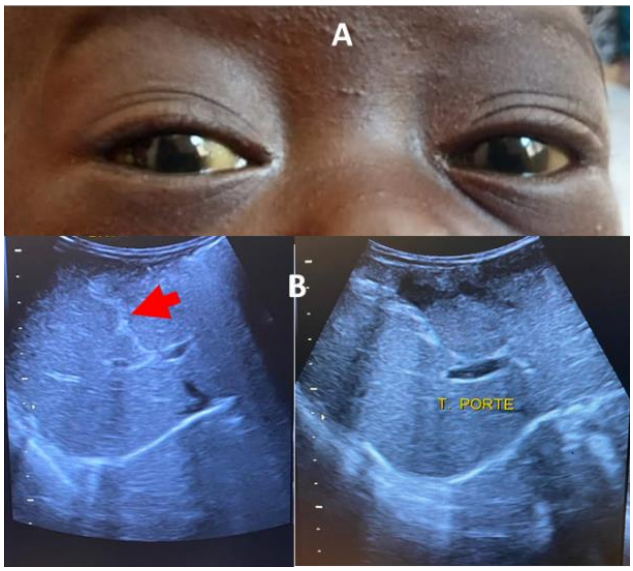

Figure 2: Photograph of a 3-month-old male infant showing frank jaundice (A) Liver ultrasound of the infant showing biliary fibrosis (red arrow), (B)

DISCUSSION

Sociodemographic data:

Frequency:

The incidence of biliary atresia varies from 5/1,000 live births in the Netherlands [2] to 32/100,000 live births in French Polynesia [1], with a predominance in Asia and the Pacific region [6].

There have been few studies on the subject in Mali. A study conducted in Bamako, Mali by M. Aremu Ya et al [5] in 2020 in the Pediatric Surgery Department of Gabriel Toure University Hospital, found a hospital frequency of 0.25% with a prevalence of 3 cases/year.

Gender

In our study, the female sex predominated with a ratio of 1.66. This same female predominance was found in Mr. Aremu Ysuf [5] with a ratio of 1.66.

The literature also reports a female predominance in numerous studies [7, 8, 9, 10], whereas Ndao C. L. [6] noted a male predominance with 53.33% of cases.

Age

Prenatal diagnosis of AVB remains exceptional [5]. It can be suspected based on certain ultrasound images. The presence of a cystic structure in the liver hilum, the absence of a gallbladder, or the absence of visualization of a normal gallbladder requires a postnatal assessment [8]. Prenatal diagnosis of AVB was not made in any of our patients. The average age of our patients was 45.5 days. Mr. Aremu Ya reported a mean age of 140.4 + or - 50.9 days with extremes of 23 and 210 days [5]. This difference could be explained by our small sample.

Clinical Features

AVB should be suspected in any neonatal cholestatic jaundice persisting beyond two weeks of age. Discolored stools and dark urine may develop gradually or later. Hemorrhagic signs, sometimes in the form of intracranial hemorrhage (ICH), may be indicative [8]. Jaundice was the most consistent sign in our series, as were discolored stools.

Ultrasound Features

In our study, the ultrasound was performed after a 2- to 4-hour fast by a radiologist, and a second opinion was then requested. Liver ultrasound was performed after a strict 6- to 12-hour fast (the child was on a drip) by an experienced radiologist ; it did not reveal any dilation of the bile ducts [9]. AVB can be suspected if the gallbladder is atrophic despite prolonged fasting, if it does not contract after bottle feeding, if there is a cyst in the hilum of the liver, if the elements of polysplenia syndrome are identified : multiple spleens, preduodenal portal vein, absence of retrohepatic inferior vena cava, median liver, abdominal situs inversus [9].

Gallbladder

Biliary tract abnormalities were the main indicators for identifying biliary atresia. These abnormalities included the absence of gallbladder, small gallbladder size, abnormal gallbladder shape and wall, and no gallbladder contraction. Abnormal wall features included wall irregularity and a thinner wall without mucosa.

The gallbladder was identified in 25%, or 2 patients, with peri-gallbladder atrophy and fibrosis. The gallbladder lumen was interrupted in 1 case, with failure of the gallbladder to empty after feeding. A gallbladder size ≤ 19 mm is a strong positive argument for a positive diagnosis of gallbladder atresia [10]. Irregularity of gallbladder wall contours is an ultrasound sign with a sensitivity of 74.23% and a specificity of 93.81% for the detection of AVB and therefore appears to be a marker with a high diagnostic capacity [10]. In our case, the gallbladder walls were thick and irregular with fibrosis.

The Cord Sign

Periportal hyperechogenicity (or “triangular cord sign”) is a very specific sign: its presence strongly supports the diagnosis of AVB (Sp=0.98; LR+=38). Its presence thus constitutes a very discriminating diagnostic element for the diagnosis of AVB, which is consistent with the literature [11, 12, 13]. Nevertheless, it is a less sensitive ultrasound sign than the signs previously described (Se=0.78) [10]. This is explained by the fact that this sign is progressive, and therefore, uncommon if the diagnosis is early [10]. Indeed, the pathophysiological hypothesis suggests that this sign would be linked to secondary periportal sequelae, explaining its later appearance. This is emphasized by Hwang *et al.,*[14], which showed that the triangular cord sign was present in 17% of children under 30 days old and 56% of the older group. We found the cord sign in 37.5% of our patients. The cord sign had a sensitivity of 100% and a specificity of 75% in the diagnosis of gallbladder atresia in Ndao C L [6] this could explain the diagnostic delay in Africa.

Cysts And Microcysts

Intrahepatic biliary cysts are rarely observed in patients with BVA. They manifest as multiple intrahepatic biliary cysts (biliary lakes) [15].

In our study, they were present in two patients, or 25% of cases. Ndao C L [6] reported a 6.6% incidence of cysts and microcysts in his study.

Splenic Abnormalities

The syndromic form of BVA is associated with splenic abnormalities (mainly polysplenia, but also splenomegaly or asplenia), intestinal malrotation, portal and caval venous abnormalities, and aberrant hepatic arteries. Humphrey and Stringer [13], as well as Koob *et al.,*[16] reported that polysplenia in the context of neonatal cholestasis was associated with low sensitivity

but high specificity for the diagnosis of AVB. These findings were also observed by C. Chardot *et al.,* [9], with a sensitivity not exceeding 26.8% and a specificity approaching 91.8%, which remains consistent with Humprhey and Stringer, who reported a low sensitivity of 10% and an excellent specificity of 100% for polysplenia [13]. In our study, we had one case of polysplenia; this infant also had cirrhosis.

Therapeutic and Progression Aspects

All our patients underwent surgery; they underwent a hepatoportoenterostomy (Kassai procedure), with favorable outcomes in six patients, and two deaths. The earlier the Kasai procedure is performed (without considering the classic threshold values at 45 or 60 days of life), the better the child's chances of being alive with their native liver in adolescence [17].

CONCLUSION

Biliary atresia is a rare condition. Prenatal diagnosis is exceptional and remains a challenge in our region. Clinical, biological, and imaging are the cornerstones of diagnosis; imaging relies primarily on ultrasound in our context after birth. The treatment is surgical and the prognosis depends on the early treatment.

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