

The Outcomes and Success Rates of Kasai Operation for Biliary Atresia Cases Using Presurgical Hepatobiliary Scan: Observations at NINMAS

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ABSTRACT

Objective: The primary goal of this research is to validate the effectiveness of hepatobiliary scintigraphy (^{99m}Tc-HIDA Scan) in assessing the outcomes and success rate of the Kasai operation for Biliary Atresia (BA) cases at the National Institute of Nuclear Medicine & Allied Sciences (NINMAS).

Methods: A total of 150 patients were included in this retrospective study who were referred to NINMAS for a ^{99m}Tc-HIDA scan between January 2022 and December 2022. Comprehensive evaluations, including medical history, physical examinations, liver function tests, ultrasonography, hepatobiliary scintigraphy, and liver biopsy, were conducted for the diagnosis of biliary atresia. Among them, 16 patients had prior Kasai operations and underwent follow-up HIDA scans.

Result: Out of the 150 patients, 103 were male and 47 were female. The average age was 8.33±2.34 months. Age distribution category divided 77 patients belong to 0 to 2 months, 45 patients aged 3 to 5 months, 18 patients aged 6 to 11 months, and 9 were of more than 1 year. Following HIDA Scan, 35 patients exhibited bowel activity within 2 hours, indicating patent biliary channels. Conversely, the remaining 115 patients were diagnosed with biliary atresia as their bowel activity was not observed within 2 hours during the HIDA Scan. Of these 115 patients, 65 were also experiencing hepatic insufficiency, with 49 of them falling within the 0 to 3-month age range. Additionally, among the positively identified cases, 16 patients had undergone the Kasai operation for follow-up, all within the 0 to 2-month age range. Post-Kasai operation, 15 patients displayed positive outcomes, while only one patient, older than 6 months, experienced a recurrence of biliary atresia, which was verified through HIDA Scan.

Conclusion: This study underscores the significance of age in effectively managing Biliary Atresia. Performing the Kasai procedure within the first 60 days after birth is associated with improved outcomes, as demonstrated by the findings of this research.

Keywords: ^{99m}Tc-HIDA Scan, Biliary Atresia, Hepatobiliary Scintigraphy.

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INTRODUCTION

Neonatal cholestatic jaundice is a condition characterized by elevated levels of conjugated bilirubin, indicating potential liver or bile duct issues. Significant elevation occurs when the conjugated bilirubin surpasses 1 mg/dl and the total serum bilirubin is either 5 mg/dl or lower, or it surpasses 20% of the total when it exceeds 5 mg/dl (1). The occurrence of this condition is estimated to be 1 in 2500 live births, according to the recent guidelines for evaluating jaundice in newborns (2). Neonatal cholestasis, often a pathological sign, typically points to underlying hepatobiliary disorders, with biliary atresia (BA) and neonatal hepatitis (NH) emerging as the primary culprits.

BA is a progressive cholangiopathy characterized by inflammation and fibrosis affecting the extrahepatic biliary system. This leads to the obstruction of bile flow due to the narrowing or closure of the ducts responsible for transporting bile from the liver to the gallbladder. Initially considered a congenital malformation, BA involves a developmental abnormality in the bile ducts, whether inside or outside the liver. In infants, BA stands out as one of the most complex liver diseases, demanding a prompt diagnosis to optimize outcomes. The challenging nature of this condition underscores the importance of early identification for effective management and improved prognosis in affected infants.

PATIENTS AND METHODS

This retrospective study included 150 referred patients (103 male and 47 female) of suspected biliary atresia who

were referred to NINMAS for biliary scintigraphy with ^{99m}Tc-HIDA Scan assessments studied between January 2022 to December 2022. Categorically the patients were grouped according to age ranging from 0 to 2 months (77 patients, 51.3%), 3 to 5 months (48 patients, 32%), 6 to 11 months (18 patients, 12%), and over 1 year (9 patients, 06%), with a mean age of 8.33 ± 2.34 months. Prior to the study, institutional ethical clearance was obtained, ensuring adherence to ethical guidelines. Additionally, informed consent was diligently obtained from the guardians or parents of the participating infants.

A thorough diagnostic approach was undertaken, involving meticulous reviews of medical history, comprehensive physical examinations, liver function tests, ultrasonography, hepatobiliary scintigraphy, and liver biopsy, to identify cases of biliary atresia (BA).

Patients were administered an intravenous injection of 2-3 mCi (0.05 mCi/Kg body weight) of ^{99m}Tc- Mebrofenin. Imaging was done with a single-head gamma camera equipped with a high-resolution collimator and patients positioned supine. Serial anterior static images were captured at 5 minutes, 15 minutes, 30 minutes, 45 minutes, and hourly for 2 hours. Delayed image at 24 hours were taken in cases of absent tracer uptake. Images were analyzed by Nuclear Medicine physicians and positively reported as BA, if images show robust intense liver uptake without intestinal activity within 24 hours.

RESULT

Total 150 patients (M= 103, F= 47) with a male to female ratio of 2.2:1 and a mean age of 8.33 ± 2.34 months and their age distribution are shown in Figure 1 and Figure 2.

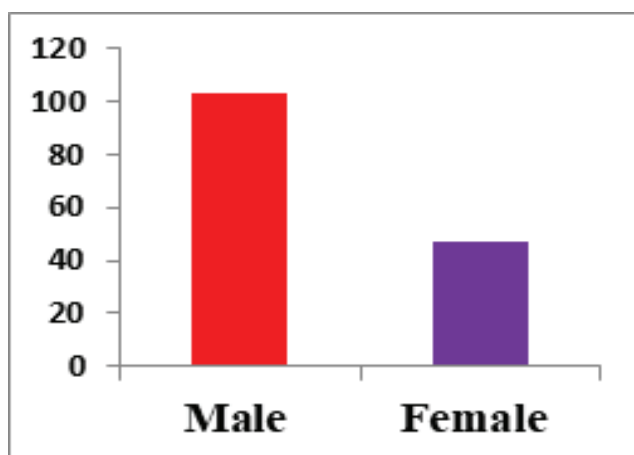


Figure 1

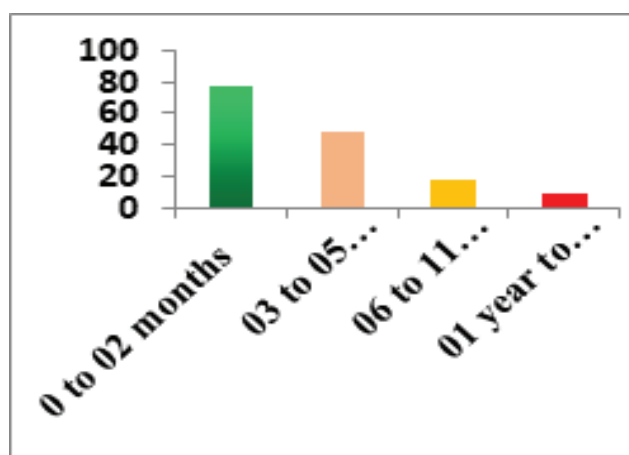


Figure 2

Figures 1 and 2 Among a total of 150 patients studied with ^{99m}Tc-HIDA, there was 1. a male-female ratio of 2.2:1; and 2. There are four categories according to age: 77 patients aged 0 to 2 months, 45 patients aged 3 to 5 months, 18 patients aged 6 to 11 months, and 9 patients aged more than 1 year.

Most of the patients were suffering from cholestatic jaundice 98 (65.3%) . In addition, 65 (43.3%) patients had contracted gallbladder, 53 (35.3%) had normal gallbladder

and 32 (21.3%) did not have one at all. The TORCH test findings were negative in 9 (6%) cases and positive in 25 (16.6%). We found 27 (18%) patients with negative Cytomegalovirus (CMV) and 61 (40.6%) patients with positive CMV. (Table 1).

Table 1: Ultrasound, TORCH and CMV Results

USG findings of GB			TORCH		CMV	
Contracted GB	Normal GB	Absent GB	Positive	Negative	Positive	Negative
65 (43.3%)	53 (35.3%)	32 (21.3%)	25 (16.6%)	9 (6%)	61 (40.6%)	27 (18%)

The latter group exhibited significantly elevated levels of total bilirubin, direct bilirubin, and alkaline phosphatase with a mean serum bilirubin level of 10.35mg/dl. Out of 150 patients, only 8 had a histopathological report, revealing Biliary atresia in their findings.

After a HIDA scan, bowel activity was positive within 2 hours in 35 (23.3%) patients indicating patent biliary channels. Meanwhile, delayed image at 24 hours showed no tracer activity, indicating no bile excretion in 115 (76.6%) out of 150 patients and confirming the diagnosis of Biliary atresia (Figure 3 and 4)

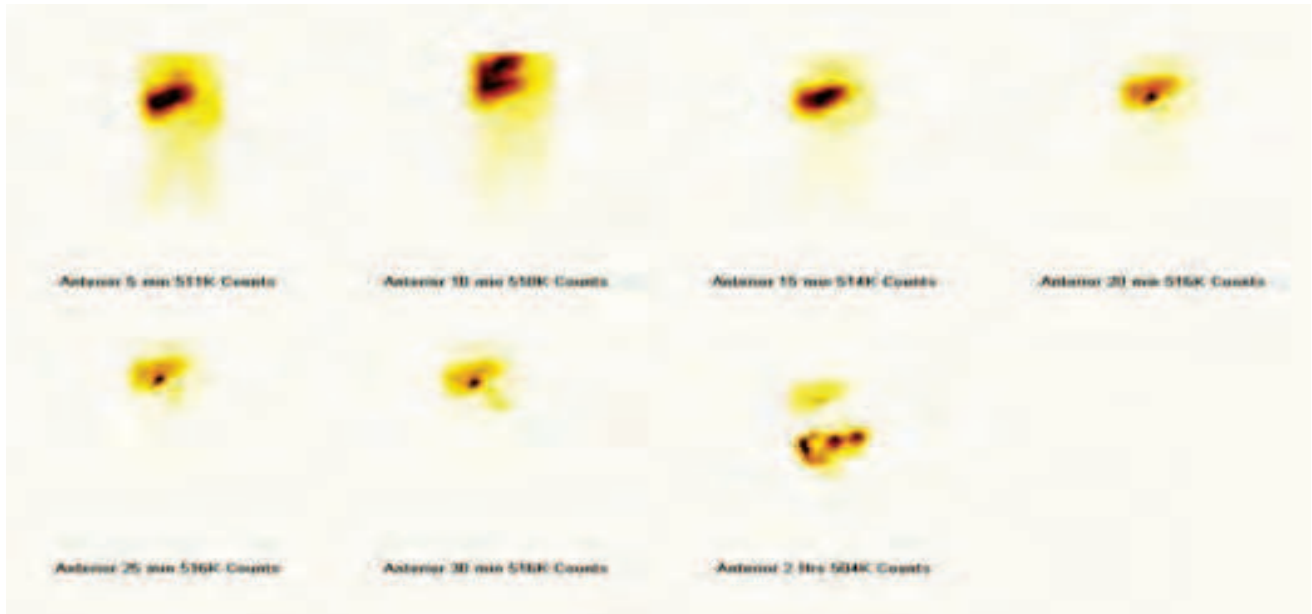


Figure 3: ^{99m}Tc-HIDA scan image of a 3months old boy with history of neonatal jaundice showing patent hepatobiliary system



Figure 4: ^{99m}Tc-HIDA scan image of a 2months old boy with history of cholestatic jaundice showing positive hepatobiliary scan.

A total of 65 (56.5%) out of 115 patients were affected by hepatic insufficiency, with a significant portion falling within the 0-3 months age range. In all the positively identified cases, 16 (13.9%) patients underwent the Kasai operation for follow-up, with their age range predominantly falling between 0 and 2 months. Following the Kasai operation, positive outcomes were observed. Only one patient of about 6 months age, encountered a recurrence of biliary atresia and confirmed by HIDA scan.

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Table 2: Response of Kasai operation among the study subjects who underwent surgery (n=16)

After KASAI Operation	
Patent	Recurrence Biliary Atresia
15 (93.75%)	1 (6.25%)

DISCUSSION

In this study, 76.66% of patients tested positive for HIDA, indicating a biliary blockage, while 23.3% tested negative. Though there are numerous causes of chronic conjugated hyperbilirubinemia in neonates, BA and neonatal hepatitis (resulting from Cytomegalovirus, Hepatitis A virus, Hepatitis B virus, Rubella, and Toxoplasma infection) account for 70-80% (3).

Positive Cytomegalovirus cases were 40.66% with 16.66% TORCH positive. Persistent direct hyperbilirubinemia and icterus typically manifest within 1-2 months of age. In the study by Abou-Taleb et al. and Dong et al. (4, 5), it was observed that biliary atresia (BA) tends to be diagnosed at a younger age, with patients being identified at approximately 8.33 ± 2.34 months. The male-to-female ratio in the current study was found to be 2.2:1, which contrasts with the findings of a previous study conducted at NINMAS by Nasreen et al (6). In Nasreen et al.'s study, it was observed that both sexes were almost equally represented in the study population.

^{99m}Tc trimethyl bromo iminodiacetic acid was preferred for hepato-biliary scintigraphy due to its high 98% hepatic extraction and minimal 1.5% urinary excretion.

(7). In neonates, mebrofenin is promptly extracted by the liver, achieving uniform distribution within 5 minutes. The gallbladder becomes visible as early as 10 minutes, and bowel activity is observed between 30 and 40 minutes. Through the many causes of persistent cholestatic jaundice in infancy, BA and Neonatal hepatitis account for 70–80% of cases (6).

BA constitutes a surgical emergency, and the ideal time for the Kasai Portoenterostomy surgical treatment is within the first 60 days of life. Failure to undergo surgery within 90 days may lead to progression into liver cirrhosis and hepatic failure, making recovery to a normal condition challenging (8). Conversely, conservative treatment proves highly effective in managing nearly all cases of neonatal hepatitis (4). No preoperative diagnostic algorithm has demonstrated an absolute diagnostic precision of 100% for biliary atresia (9). Out of 115 patients, eight were diagnosed with BA by histopathological examination, while the other 107 cases were diagnosed using hepatobiliary scintigraphy only.

This study included those 16 patients (10.66%) who underwent follow-up after the Kasai operation and among them only 1 patient (6.2%), older than 6 months, showed non-patency of the biliary tree. These findings underscore the critical window for surgical intervention

in the context of biliary atresia and highlight the significance of timely follow-up for optimal patient outcomes.

CONCLUSION

The study highlights the importance of age in managing Biliary Atresia (BA), a leading cause of liver transplant in newborns. Early diagnosis with biliary scintigraphy followed by intervention, particularly Kasai operation within the first 60 days after birth is crucial for timely intervention and improved outcomes.

Conflict of Interest: The authors have no conflicts of interest.

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