*Bahrain Medical Bulletin, Vol. 36, No. 2, June 2014*

# Cystic Biliary Atresia: Why Is It Important to Distinguish this from Congenital Choledochal Cyst?

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## ABSTRACT

**We present a 2-month-old jaundiced infant with a rare type of biliary atresia who appeared to have a choledochal cyst on magnetic resonance cholangiopancreatography (MRCP) and ultrasound. Intra-operative findings were the only proof of biliary atresia. Following portoenterostomy, the liver function tests (LFTs) and bilirubin levels were returned within normal range and his jaundice was resolved.**

**Any neonate presenting to a pediatric clinic with prolonged jaundice lasting more than two weeks, especially in cases of direct hyperbilirubinemia, must be thoroughly assessed and referred as early as possible for a pediatric surgical opinion to rule out the possibility of biliary atresia.**

***Bahrain Med Bull 2014; 36(2):105-107***

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## INTRODUCTION

Neonatal jaundice is a common problem and in the majority of such infants the elevated bilirubin is a physiological consequence of immature conjugation. While such "benign" jaundice usually clears within the first 3-4 weeks of life, jaundice extending beyond the 4$^{th}$ week is considered pathological and worthy of further investigation and possibly intervention. Thorough investigation is mandatory at this stage as the consequences of missing a correctable surgical cause could have significant effects. Every effort is made to distinguish non-surgical and surgical jaundice, especially Biliary Atresia, as the outcome following portoenterostomy for Biliary Atresia (BA) is significantly better when surgery is performed under 6 weeks. Early referral to a pediatric surgeon is therefore essential to optimize outcome especially when a possible diagnosis may be biliary atresia[1].

The aim of this report is to present a case of a two-month-old child with a history of intermittent yellow and pale stool and a cystic dilatation at the porta hepatis.

**THE CASE**

A two-month-old male presented to the pediatric clinic with a history of prolonged jaundice from the third day of life and persisting to the time of presentation. The stool was intermittently yellow and pale in nature and in spite of normal feeding and activity the neonate failed to thrive. Laboratory investigations revealed an elevated conjugated bilirubin of 113 umol/L. An ultrasound showed cystic structure at the porta hepatis, a normal gallbladder consistent with a choledochal cyst. A severely jaundiced child weighing 4.3 kg with mild liver enlargement was examined. Hepatobiliary Iminodiacetic Acid scan was not available and magnetic resonance cholangiopancreatography revealed a cystic dilatation at porta hepatis, normal gallbladder but no common bile duct suggestive of a choledochal cyst.

The history and magnetic resonance cholangiopancreatography suggested the need for urgent laparotomy with or without on-table cholangiogram, see figure 1a, b. Intraoperative findings included an atretic gallbladder with a cystic dilatation at the porta hepatis and no common bile duct confirming a diagnosis of biliary atresia, see figure 2. A standard Kasai porto-jejunostomy was performed and a wedge biopsy was taken from the liver. Histopathology of the portal plate confirmed fibrosis, absence of the normal ductal structures, several microscopic canaliculi, bile duct plugging, duct proliferation, portal tract expansion and some fibrosis with minimal bridging fibrosis, which confirmed the diagnosis of biliary atresia, see figure 3.

Drainage was successful as indicated by the passage of normally pigmented stools and the progressive and rapid reduction in serum bilirubin, from 138 to 65. At the time of surgical confirmation of biliary atresia, he was given steroids and continued according to Alabama protocol. At the time of discharge, he was feeding well, the weight had increased, the stool was normally pigmented and the liver function tests were improving. The patient was discharged one week after surgery; follow-up in the clinic three weeks later showed that his liver function tests were near normal.

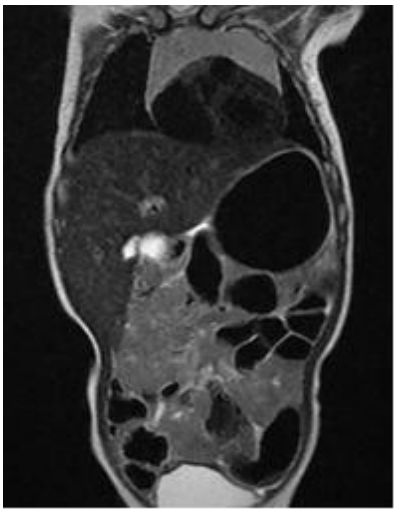


**Figure 1a: MRCP Sagittal Section**

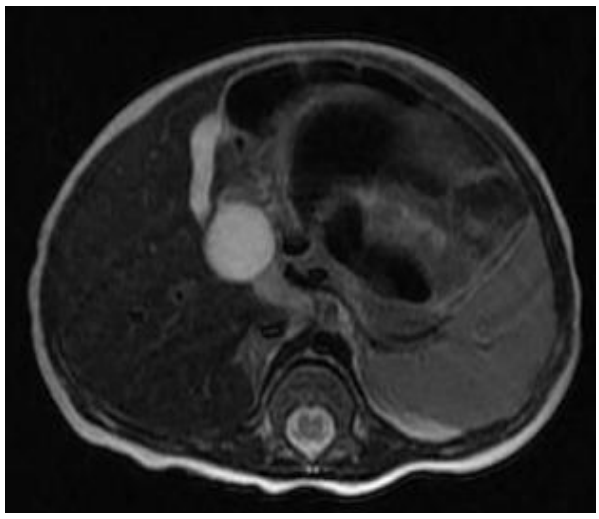


**Figure 1b: MRCP Coronal Section**

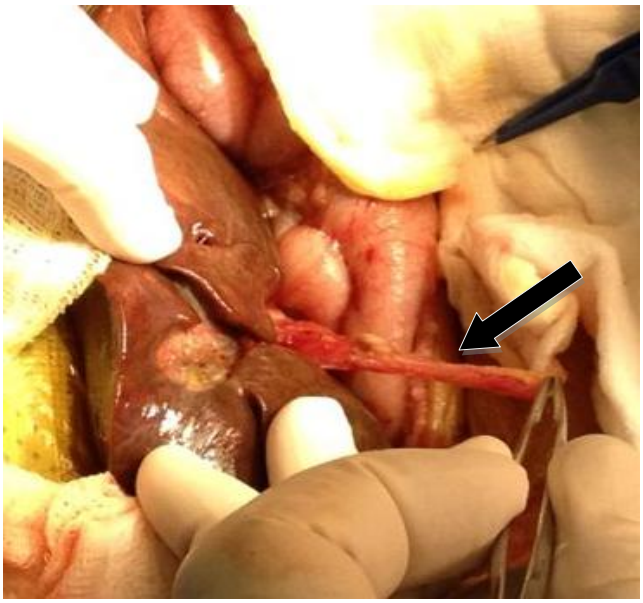


**Figure 2: Atretic Gall Bladder (Arrow) and Fibrosed Extrahepatic Bile Ducts**

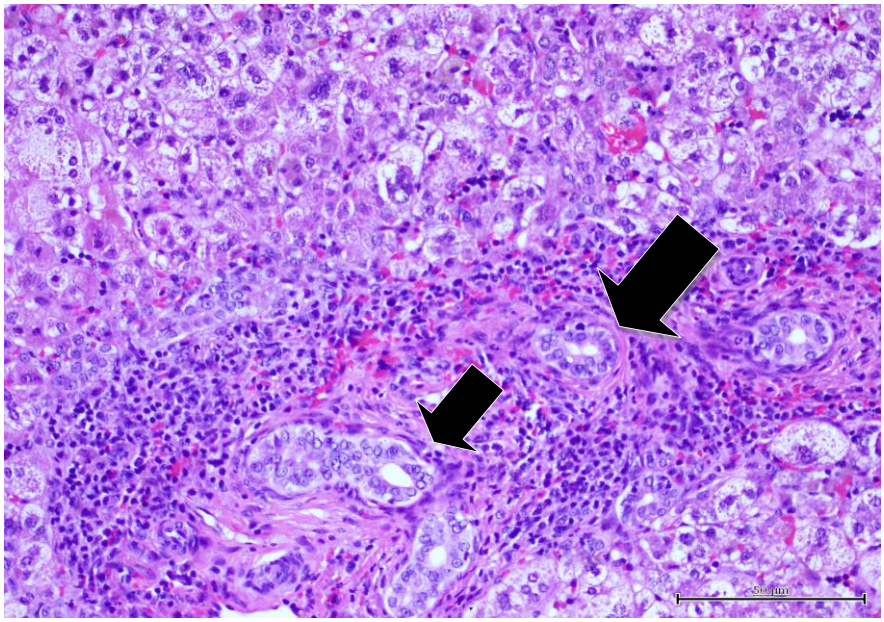


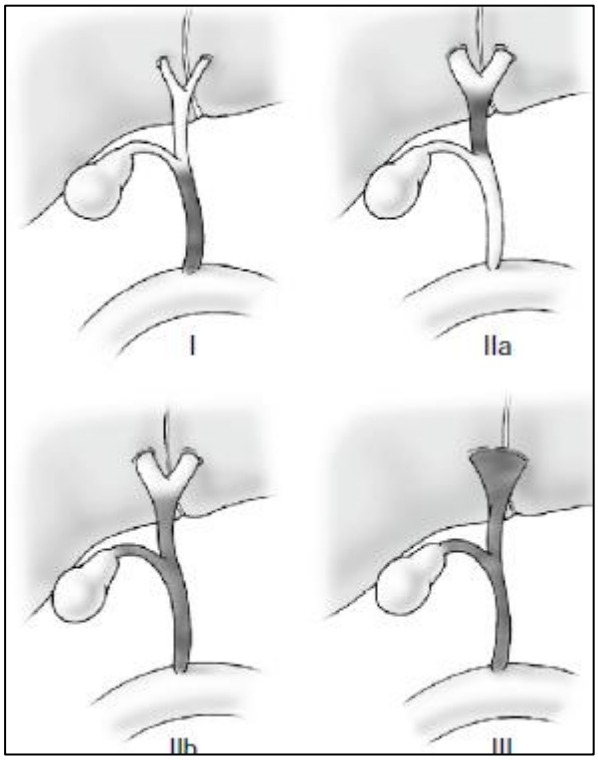
**Figure 3*:* Histopathology of Liver Portal Plate Showing Proliferating Canaliculi Surrounded By Area of Fibrosis (Arrows)**

## DISCUSSION

Biliary atresia may present with similar clinical findings as in choledochal cyst and the window of surgical intervention can easily be missed due to difficulty in the interpretation of current diagnostic methods. Biliary atresia is a rare condition (1:18,000 live births) which causes fibrosis and obliteration of intrahepatic and extrahepatic biliary ducts[2]. The clinical

presentation ranges from white chalky stool and dark urine to jaundice and hepatomegaly; although, some may have a history of slight stool pigmentation. Untreated biliary atresia leads to progressive obstruction, fibrosis, cirrhosis and finally liver failure.

One of the common reasons of misdiagnosing biliary atresia is the different types, which ranges from types I to III. Type I involves occlusion of the common bile duct; type IIa involves the obliteration of the common hepatic duct only, whereas type IIb includes the CBD, the common hepatic duct and cystic ducts with cystic dilatation of the ducts at the porta hepatis but without gallbladder involvement; type III is described as obliteration of the common, hepatic, and cystic ducts without anastomosable ducts at porta hepatis. On the other hand, choledochal cysts are classified into types A to F and could present with similar findings but at any age group. Despite ultrasound and MRCP, the two conditions can easily be confused, especially with biliary atresia type IIb and choledochal cysts type A and B, see figure 4 a, b[3].



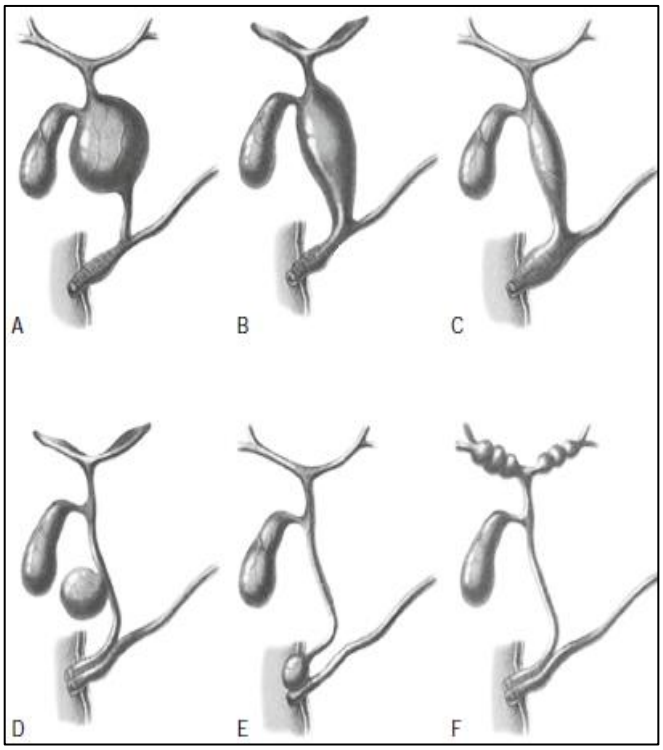




**Figure 4a: Classification of Biliary Atresia** **Figure 4b: Classification of Choledochal Cyst**

The initial presentation of this patient was with prolonged jaundice but the stools were not consistently acholic and reported to be pigmented from time to time. This history is not uncommon in choledochal cyst and combined with the initial ultrasound could cause a false sense of security and result in a delay in diagnosis and intervention with serious consequences. Fortunately in this patient, an MRCP raised serious concern and culminated in a laparotomy and successful Kasai. A diagnosis of choledochal cyst especially against the background of conjugated hyperbilirubinemia must always be confirmed by MRCP and surgical exploration performed if any doubt exists to rule out cystic biliary atresia. Ultrasound guided liver biopsy would provide the diagnosis of biliary atresia; it was not done in this patient. The threats lie in the time management of the case, whereas a choledochal cyst

can be conservatively managed and surgically treated at a later age (if not obstructive), biliary atresia must be surgically treated within a time period of 60 days.

Several studies showed the increased rate of liver transplant in patients with delayed correction of biliary atresia because time is directly proportional to the extent of liver fibrosis and cirrhosis[4]. Broad spectrum prophylactic antibiotics with good gram-negative coverage are essential in the postoperative management as cholangitis is the most common complication post Kasai[5]. High dose steroids are also used to improve bile drainage and to reduce fibrosis at the intrahepatic biliary radicles[6]. The role of steroids in improving the outcome of biliary atresia post Kasai is controversial. Recent study compared the outcome of infants with biliary atresia post Kasai who had high dose of steroids to infants with low dose of steroids and no steroids; they concluded that high dose steroids improved the outcome of infants post Kasai[7].

## CONCLUSION

**Any neonate presenting to a pediatric clinic for prolonged jaundice lasting more than two weeks, especially in cases of direct hyperbilirubinemia, must be thoroughly assessed and referred as early as possible for a pediatric surgical opinion to rule out the possibility of biliary atresia. These neonates with clinical and laboratory evidence of obstructive jaundice need urgent surgical exploration of the biliary tree with or without intra operative cholangiogram. Despite radiological diagnosis of choledochal cyst, it is imperative to rule out biliary atresia. The window of surgical treatment should not be missed as time has a significant impact on prognosis.**

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**Author contribution:** All authors share equal effort contribution towards (1) substantial contributions to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of the manuscript version to be published. Yes.

**Potential conflicts of interest**: None.

**Competing interest:** None. **Sponsorship:** None.

**Submission date**: 16 January 2014 . **Acceptance date**: 31 March 2014.

**Ethical approval:** Approved by Research and Ethics Committee, King Hamad University Hospital, Bahrain.

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