

Case Report

A Case of Intrahepatic Biliary Atresia in Pediatrics

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Received: 05 May 2021; **Accepted:** 17 May 2021; **Published:** 26 May 2021

Citation: Srikanth Darisi. A Case of Intrahepatic Biliary Atresia in Pediatrics. *Journal of Pediatrics, Perinatology and Child Health* 5 (2021): 130-135.

Abstract

The chief objective of this case report is to give an overview of how Biliary atresia (BA) occurs, presents, and management. Biliary atresia is a congenital abnormality with mostly affects the biliary tract (intrahepatic or extrahepatic) and often presents with symptoms of jaundice, the prevalence of this disease is more in Asian females than males. This case demonstrates a male patient of 6 months 8 days old he presented with fever, abdominal distension, severe anemia, and also yellowish discoloration of skin sclera and urine, he was diagnosed with intrahepatic biliary atresia, It is important to diagnose if it is intra hepatic or extrahepatic biliary atresia as the treatment varies for both, Treatment in case of extrahepatic biliary atresia is Kasai procedure but the diagnosis in his case is intrahepatic biliary atresia

Kasai procedure is not done, the patient is given symptomatic treatment and is enlisted for a liver transplant.

Keywords: Biliary Atresia; Chronic Liver Disease; Liver Transplant; Kasai Procedure; Pediatrics

1. Introduction

Biliary atresia is a congenital malformation of the parts of the Biliary tract [1] or sometimes even absent, it is commonly caused due to CMV [2] mediated inflammation and also due to autoimmune-mediated bile duct destruction [3] and there is also a role of genetic mutation CFC1 gene; if left untreated it may even cause the cirrhosis and even death of liver cells [3] it is an inflammatory disorder which can involve any part the bile tree; from the removal of

bile in the liver through intra-hepatic bile duct till the entry of bile into the intestine with the help of common bile duct, The incidence of biliary atresia is high in Asia and also the Pacific region; females are more prone to this disease more than males, it is also called as Newborn jaundice - biliary atresia.

2. Case Presentation

A 6 months male patient of 7.25Kgs was admitted to hospital with the chief complaints of abdominal distension, fever on and off since 4 months, Pale stools, yellowish discoloration of body, sclera, urine, decreased oral intake, sleep, irritability since 4 months, up on physical examination of the abdomen, patient was found to have firm tenderness, hepatomegaly, Splenomegaly, umbilical hernia, Vitals on admission were as follows heart rate of 140bpm, Respiratory rate of 25cpm, temp of 99.8°F, Spo2 of >95% Laboratory values revealed altered liver function test (T.Bilirubin- 12.14, D.Bilirubin-9.64mg/dL, Total protein of 5g/Dl, albumin -1.8g/dL, SGOT-151 IU/L, SGPT- 70IU/L), altered coagulation panel (PT-13.9 sec, INR-1.36), Total counts were elevated (25,100), Hb of 6.5g/dL, abnormal in Iron, USG abdomen showed Hepatomegaly, splenomegaly and contracted gall bladder, 2D echo was normal, Color doppler of liver was done : Non visualized main portal vein, small sized hepatic vein with monophasic flow, elevated PSV in hepatic artery with diastolic flow reversal, multiple cysts of variable size along the intra and hepatic biliary radicals, ascites, for further evaluation CT Abdomen was done in which multiple well defined hypodense cystic lesions of varying size in the region of intrahepatic biliary radicals with central dot sign was seen, patient was diagnosed to have intrahepatic biliary atresia with decompensated liver disease, He was started on Inj. Vitamin K, Inj.Piptaz, Inj, Amikacin, Vitamin E, Vitamin A, Vitamin D3, Tab.

Spirolactone, Furosemide, Syp, UDCA, SYP. Shelcal, Tab Vangancyclovir and is listed for a liver transplant.

3. Discussion

Classification of Biliary atresia

1 Based on the area affected [4] (Figure 1)

It can be classified into different types such as TYPE 1 here common bile duct is being affected, TYPE2 it is of 2 types: 2a in this type common hepatic duct is affected, 2b in this type common hepatic duct, cystic duct, common bile duct are affected, TYPE 3 this type of biliary atresia involves entire bile pathway which includes intrahepatic duct, common hepatic duct, cystic duct, and common bile duct.

2 Based on the time of onset of disease: if symptoms appear within 2 weeks of birth it is called early-onset Biliary atresia, if symptoms appear after 2 weeks of birth it is said to be late-onset Biliary atresia [5, 6].

3 Based on the organs involved

- SyndromicBA: - this type of BA is associated with various congenital abnormalities such as Asplenia, Polysplenia, cardiac or other inter-abdominal defects such as situs inversus, pre-duodenal portal vein, the absence of retro hepatic inferior vena cava, intestinal malrotation.
- Non-Syndromic BA: - It accounts for about 90% of the cases in which there is only a BA.

Patients with Biliary atresia usually present with complaints of Jaundice within the second or third week of the birth which is characterized by the yellowish discoloration of skin, sclera, and urine, Alcoholic stools which is a result of less bile in the GIT due to

obstruction or inflammation of the bile duct, infants may also have abdominal distension, liver enlargement the patient may have developed symptoms which include poor weight gain, irritability, and increase in blood pressure within the portal veins by the age of 6 to 10 weeks.

3.1 Screening of biliary atresia

Every case of jaundice that is persistent for more than 2 weeks should be checked for the Biliary atresia.

- Ultrasound: This should be done after 12 hours of fasting, if the gallbladder is shrunk despite fasting it can be suspected to be biliary atresia if there is any cyst in the liver hilum [6].
- Cholangiography: This test is performed when ultrasound is clear, to assess the morphology and patency of the Biliary Tree [6].
- Liver Biopsy: This can reveal the histological changes that occurred due to BA which include Bile plugs, Ductular proliferation, portal edema,

and also fibrosis [6].

It can also be caused by the CMV virus which can be diagnosed through the testing of antigen for CMV such as IGM and IGG.

Biochemical Liver Function Tests which include Abnormal LFT's are seen In all cases of BA, Elevation in serum Bilirubin and SGOT, SGPT, ALP; Decrease in the serum protein is seen, alterations in LFT's does not indicate the duration of disease but they reveal the extent of parenchymal damage [1]. This helps us to know if liver damage is present.

- Stool Color Cards [5]: Infant screening for BA is done by using Stool color cards in some countries, in which the color of the stool is monitored for about one month and is compared with color cards for diagnosis; the specificity of this test is low and the efficacy of this method is not yet proved As in Table No. 1.

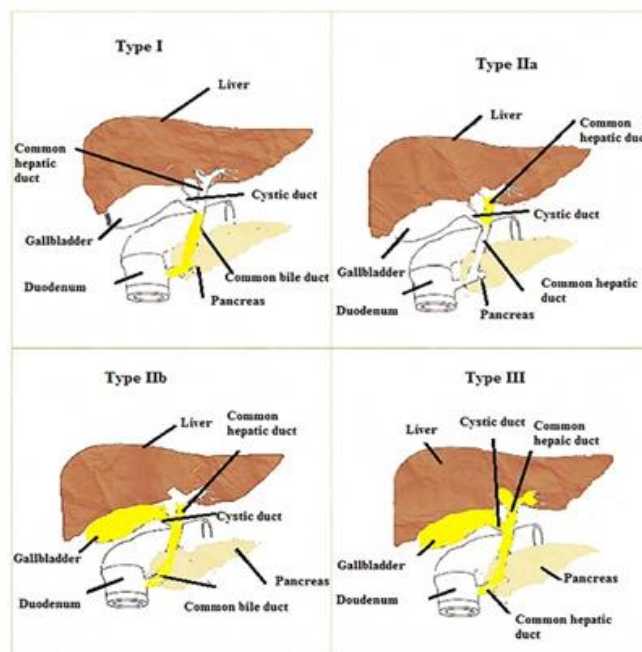


Figure 1: Classification of Biliary atresia based on the affected area.

Stool color	Remarks
Red	Bleeding lower gut or rectum
White, Clay color.	Antidiarrheal, lack of bile
Green	Antibiotics, bacterial infection
Yellow	Excess fat, small intestine infection
Black(or) dark brown	Iron supplements, bismuth sub salicylate, Bleeding in the upper digestive tract

Table 1: Stool color and remarks.

3.2 Management

During the treatment, some common medications given to the patients include multivitamins mostly Fat-soluble vitamins as there will be a deficiency of this fat-soluble vitamin due to the absence of bile [7], other medications can include Lactulose, antiviral therapy (If the BA is caused due to CMV virus), Biliary atresia is the most common indication for liver transplant in newborn; if diagnosed early it can be treated with surgical intervention called a KASAI procedure also called hepatoportoenterostomy before the development of this KASAI procedure, patients with BA had a very low prognosis rate [3]. If diagnosed early i.e. within 60 days of birth a hepatoportoenterostomy can be performed, in this procedure the necrotic part of the bile duct is surgically removed and the healthy duct is connected to the intestine for the flow of the bile [8] and a need of liver transplant can be minimized; but the success of this procedure depends upon the early diagnosis of the disease and this procedure is referred to as KASAI procedure and is only done if the child age is less than 60 days, however in some cases the patient may need the liver transplant within 2 years after this procedure Common complications of this procedure include infection and failure of this procedure [9], If the KASAI procedure fails to restore the bile flow from the liver to the intestine the alternative treatment option which are available is a

liver transplant which in most of the cases occur at the age of 2 years from the KASAI procedure and can also occur even before that if the condition is severe The success of KASAI procedure depends upon the age of performing the KASAI operation [10-13].

In the case of Intrahepatic BA, we can do a liver transplant by obtaining a liver from any cadaveric donor or from a living parent donor (The cadaveric donor is a donor who is of the same age only a few lobes of the liver are taken for this procedure), The mortality rate of patients waiting for a liver transplant has decreased in recent times due to the development of new techniques such as liver graft splitting [14], living related donor [15].

4. Conclusion

In this case reported, the cause of BA is due to inflammation which is mediated by CMV and the patient was diagnosed having BA with Caroli's disease (inflammation of intrahepatic bile ducts) and is listed for a liver transplant, The patient is started with antibiotics to help reduce the inflammation, antiviral therapy for CMV virus, other supportive measures such as multivitamins to treat the malabsorption due to decreased secretion of bile into the duodenum, According to recent research the best

nutrition treatment for BA patients before liver transplantation include cycled enteral nocturnal feeding with daily oral feeds using formula (16)(17). After liver transplant it is recommended that the patient is on parental feeding for two days for recovery, later patient can have enteral feeding.

Acknowledgment

The author(s) would like to acknowledge, Karnataka College of Pharmacy, Karnataka, India for providing an institutional research platform and necessary facilities.

Competing Interests

The author(s) declare that they have no competing interests

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