Billiary atresia: Three cases after kasai portoenterostomy

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ABSTRACT

Biliary Atresia (BA) occurs in about 1:15,000 live birth. The diagnosis is confirmed by Cholangiography. Kasai Portenterostomy (KPE) is one of the managements and age at surgery is the important factors for the success. This case report described the clinical presentation and management of BA. Three cases of BA were reported during 2016. They were two cases of two months old and four months old. All came with persisted jaundice skin > 14 days of life. The laboratory showed elevated direct bilirubin serum. Three of them underwent cholangiography and KPE. After surgery, one suffered from repeated Cholangitis, one repeated abdominal leakage, and other without complication. All of these three cases were survived until this case report was made. The diagnosis of BA should be considered for any infant who came with jaundice skin which persist after 14 days of life. Age at KPE is an important determinant of jaundice clearance.

Keywords: biliary atresia, jaundice, acholic stool, triangular cord, kasai, child


INTRODUCTION

Biliary Atresia (BA) is a progressive obstructive cholangiopathy of both intra- and extrahepatic biliary tree to various degrees, leading to obstructive bile flow, jaundice in neonates, cholestasis, and cirrhosis.¹,² It is a rare condition that occurs in about 1:15,000 live birth (which is 1.5 times much more common in females than males).³,⁴ It is an emergency condition that requires immediate surgery intervention. Its represents the most common cause of pediatric end-stage liver disease, and the leading indication for pediatric liver transplantation.⁵ If this condition is left untreated, progressive liver cirrhosis leads to death by the age of two years.¹,⁵ The treatment of BA involves a sequential strategy combining Kasai Portenterostomy (KPE) as first line and Liver Transplantation as second line treatment if KPE fails to establish bile flow and/or progressive liver fibrosis and cirrhosis occurs.²,⁶ The most significant factor correlating with success of KPE is the infant’s age at the time of surgery. Among infants who undergo the KPE procedure within 60 days of life, 57%-90% will have successful reestablishment of bile flow, but the success rate drops to <20% if the procedure is performed after 90 days of life.⁷ This time-sensitive success rate makes the infant’s age at the time of surgery crucial to optimize outcomes and decrease short-and long-term morbidity and mortality.
mortality, and underscores the importance of early
diagnosis of biliary atresia.8,9

Classic clinical manifestation in infants with
BA are icteric sclera and/or jaundice skin, hepato-
splenomegaly, and acholic stool.2 Those infants
will generally appear healthy as newborn, they do,
however, exhibit jaundice at birth or shortly there-
after and may be clinically indistinguishable from
infants with non-conjugated or indirect hyperbil-
irubinemia, such as “physiological jaundice” and
“breast milk-associated jaundice”.7 BA must be
suspected in all neonates in whom jaundice persist
after the period of physiologic hyper-bilirubinemia
(first 14 days of life). The diagnosis mostly delayed
because of difficulties in differentiating jaundice due
to liver disease from physiologic jaundice and iden-
tifying an abnormal stool color.7 Later, sequences
of para-clinical investigations such as biochemical
tests, ultrasonography, hepatobiliary scintigraphy,
liver biopsy, and ultimately intraoperative cholan-
giographies as a gold standard are employed to find
out infant with BA as soon as possible at the earliest
possible opportunity.2

In this paper we present the experience of
Department of Child Health and Pediatric Surgery
in managing some cases of Biliary Atresia with
varies outcome. The objective of this case report is
to describe the clinical presentation and manage-
ment of BA which then can improve the outcome
of any patient with Biliary Atresia.

CASE ILLUSTRATION

First case
A two months old baby girl was come to our insti-
tution with chief complaint of jaundice. She has
been reported having yellowish skin since she was
three days old. For that reason, she has had history
of having three days of phototherapy in hospital
then continue to have daily sun bathing at home. At
seventh days old, she was reported with worsening
jaundice skin, she went to pediatrician and having
two kinds of drugs which should be taken for two
weeks. On 1st of February 2016, the two phases of
abdominal ultrasonography were performed due
to persistent jaundice. It was revealed the sign of
extrahepatic cholestasis with suspected caused by
implicated bile syndrome. The patient was continued
to have the previous two drugs for another five
weeks. On 5th of April 2016, the second of two phase
abdominal ultrasonography was proceed due to
the persistence jaundice on the skin and pale stool
and also with a remarkable result of cholestasis
(total bilirubin 10.44mg/dL; conjugated bilirubin
9.36 mg/dL; Gamma GT 715 U/L(increased
23 times); SGOT 158.50; SGPT 112.20). The second
ultrasound showed the same result as the first one,
and as the recommendation from the pediatrician,
the family agreed to proceed cholangiography and
if needed will be right away proceed the Kasai
procedure as the definitive treatment. The cholan-
giography was performed on 15th April 2016,
confirmed biliary atresia and then continued with
Kasai procedure uneventful.

During the observation in hospital after the
Kasai procedure, the symptoms of jaundice was
going better, the patient then allowed to discharge
and continue treatment with oral prophylaxis
antibiotic and probiotic and others medicines as
supportive treatment such as ursodeoxycholic acid
and multivitamin. However, unfortunately, one
month after the procedure, she developed high
fever, the skin became yellowish again with pale
stool. The patient was admitted to hospital with
suspicion of cholangitis. At that moment she has
been having five days of treatment with antibiotic
Cefoperazone Sulbactam, and then discharge on
day 6 due to improving condition. Three days later,
she was admitted again in hospital due to developed
again high fever, about 39°C which then occurred
almost every day. Another symptom accompa-
nied were a jaundice skin of the body for two days
before admission. Yellowish to greenish color was
obvious on both eyes, face, chest, abdomen and
has no improvement. The pale stool was reported
for two days before the admission, along with the
jaundice skin and the tea color urine. Laboratory
result initially showed anemia (with Hemoglobin
7.14 g/dL), trombocytopenia (platelet count down
to 177.6 K/ul), normal electrolytes, then the liver
function tests revealed: total bilirubin 8.29 mg/dL
(increased 8 point), direct bilirubin 7.22 mg/dL
(increased 24 times), indirect bilirubin 1.07 mg/dL,
SGOT 61.10 U/L (increased two times), SGPT
67.0 U/L (increased two times), albumin 3.11 g/dL,
globulin 1.4 µg/d µg/dL, ALP 119 mg/dL, gamma
GT 702 U/L (increased 22 times). BUN 24 mg/dL;
creatinine 0.28 mg/dL. PT 18.2 (control 14.4 second);
INR 1.61; APTT 50.5 (control 36 second). Aspartate aminotransferase to platelet ratio index (APRI) score= 1.27 (suspected with fibrosis process). The blood culture came with positive infection of Staphylococcus Hominis sp which then confirmed the diagnose of Cholangitis (repeated case). The patient was treated with Cephalosporin 3rd generation (ceftriaxone 50 mg/kgBW every 12 hours) and steroid (Dexamethasone 0.5mg/kgBW/day divided three time per day. Ursodeoxycholic acid was given with range of dose 10-30 mg/kgBW/day. Calories received was 120-150% RDA by MCT formula. Vitamin including vitamin K injection maintenance 0.3 mg/kgBW every 3-4 weeks intramuscularly, Vitamin A 5000-25.000 unit per day, Vitamin D3 calcitriol 0.05-0.3 micro/kgBW/day and Vitamin E 25-50 international unit/kgBW/day were given right away for supportive treatment. On the day 10 of admission, she showed significant improvement, she was doing active, drank well, still jaundice and slight pale, without fever, nor acholic stool. The laboratory result including routine hematology test are: WBC 20.96 K/uL (neu 51.02%; lim 37.27%); hemoglobin 8.75 g/dL; hematocrit 29.38%; MCH 98.61 fL; MCHC 29.79 g/dL; trombocyte 560.60 K/uL. Total bilirubin was 6.62 mg/dL (increased 6 times), direct bilirubin was 6,44 mg/dL (increased 21 times), bilirubin indirect was 0.18 mg/dL (increased 6 times), direct bilirubin was 6,44 mg/dL (increased 21 times), bilirubin indirect was 0.18 mg/dL (increased 6 times), SGOT 91.30 U/L (increased three times), SGPT 141.30 U/L (increased 4 times), APRI score = 0.603 (normal).

Reviewed back from her past, she was born by spontaneous delivery with birth weight was 2100 grams (low birth weight baby) and 50 cm in length. No history of consanguinity of her parents. Mother has no history of having any infection during pregnancy. Since she was born, she was full fed with breast milk until four- month- old, but then switched to standard infant formula since then with 40-50 ml every 6-8 hours. She was having good and normal growth and development as her age. Until this study was made, she got her vaccina - some completely with catch up schedule due to her illness.

Second case
A four months old baby boy, came to our institution with chief complaint of icteric on whole body. He was found icteric with slightly greenish color of the skin since he was two days old. It was initially obvious on both of eyes, which then spreading to whole of the body and persisted. For this symptom, he has been having phototherapy when he was 2 weeks old. He also reported having pale stool when he was 1,5 months old with brownish color of the urine.

He was born by cesarean section, with birth weight was 2770 gram. No history of consanguinity of her parents. This boy is a second child in his family. His brother had no same symptoms with him. Mother had normal history of pregnancy, no history of TORCH infection nor Hepatitis B infection and none others infection. On presentation, generally his skin was looked icteric on whole of his body. The vital sign was within normal range, no dysmorphic face. There were icteric eyes, no others abnormality was found. Chest examination revealed normal. There was distention of the abdomen with enlargement of the liver and spleen without any sign of ascites. His body weight was 6.4 kgs and length was 61 cms, which showed he was well nourished.

Laboratory study showed an increasing some of the liver function test panel, it was including the total bilirubin (6.07 mg/dl, which was increased 6 points), direct bilirubin (5.33 mg/dl, which increasing 18 points), SGOT (396.5 U/L, which increasing 14 times), SGPT (283.9 U/L, which increasing 8 times), and the Gamma GT (328 U/L, which increasing 10 times). Another laboratory study showed that was slightly increasing of the Prothrombine time (13.14 second). Thyroid function revealed normal. The CMV serology showed that the IgM CMV was equivocal and positive result of the IgG anti CMV. Urine analysis was positive for leucocyte macroscopically and was found 1-2 leucocytes on the urine sedimen. No microorganism was growth on the urine culture. Stool observation showed consistent of three acholic stool appearance. From two phase of liver ultrasonography we found there was no gallbladder inflammation, but there was hyperechoic area found on the anterior edge of the portal vein wall, 4 mm in width, with positive sign of triangular chord and dilatation of the hepatic artery, 2.8 mm. With all of those data, the patient was diagnosed as extrahepatal cholestasis et cause suspect biliary atresia.

The patient then undergo cholangiography intraoperatively which then revealed there was no

Picture 2 The Kasai Portoenterostomy
biliary duct, the Kasai procedure was performed right away uneventfull and the liver tissue was sent for pathology anatomy examination. Final assessment of this patient confirmed as Extrahepatal Cholestasis et cause Biliary atresia. Post Kasai procedure, patient was observed and treated in PICU. He received antibiotic combination including Gentamycin 12.5mg/kgBW every 8 hours combined with Amoxycilin 25 mg/kg every 8 hours. He also received methylprednisolone 20 mg in day 1, continue with 2.5-5 mg/kgBW everyday for 2 weeks. Another antibiotic was given to prevent the incidence of cholangitis post Kasai procedure, it was Cefxime 5mg/kgBW every 12 hours for 1 month. Ursodeoxycholic acid was given with range of dose 10-30 mg/kgBW/day. Calories was received 120-150% RDA by MCT formula. Vitamin including vitamin K injection maintenance 0.3 mg/kgBW every 3-4 weeks intramuscularly, Vitamin A 5000-25.000 unit per day, Vitamin D3 calcitriol 0.05-0.3 micro/kgBW/day and Vitamin E 25-50 international unit/kgBW/day were given right away for supportive treatment. Observation post Kasai procedure were including clinical symptoms (icteric, fecal color, and fever), laboratory profile (total bilirubin, direct bilirubin, SGOT, SGPT, albumin, and hemostasis profile such as Prothrombine time and INR), and cholangitis sign. All of those monitoring points were evaluated every month until cholestasis improved clinically and laboratories.

Three days post Kasai procedure, patient with no improvement of icteric and patient still with abdominal distention. Plain photo abdomen showed a free air in peritoneum which impressed perforation condition. Re-open abdominal surgery was performed right away due to leakage. The patient continued observed in PICU and continued treatment. Seven days post Kasai procedure patient still without improvement of icteric and still with abdominal distention. Plain photo abdomen again revealed a suspicion of abdominal perforation. The second re-opening abdominal surgery was proceeded right away. Patient continued treatment and observation in PICU. After seventh days observation in PICU, he was moved to general ward and then discharged few days later. On discharge, family were educated about the need of routine evaluation of clinical symptoms and laboratory profile (at least every 3-4 week). Preparation for liver transplant, avoidance of any drugs consumption which can damage the liver condition.

**Third case**

A two-month-old baby girl was coming to our institution with chief complained of icteric. She was reported with yellowish to greenish color on her body since few days after birth. Icteric was initially on both eyes which then spreading to whole body since then. She had pale stool since birth and tea color of urine. She had no others past medical history, no long fasting history.

She was born by cessarian section, aterm, with birth weight 3800 gram. She is the only child of her parents. No history of consanguinity of her parents. Any icteric history on her family was denied. Mother has normal condition during pregnancy, no history of having any infection including TORCH infection, no hepatitis B infection. Others infection were denied.

Physical examination revealed that generally the patient looked icteric and the vital sign still on the normal range. No dysmorphic face. Both eyes were icteric, no others abnormality were found. No abnormality was found on chest examination. No hepatosplenomegaly. Anthropometric measurement showed she was well nourished, with body weight 5.7 kgs and height 61 cms.

Laboratory study showed an increasing some of the liver function test panel, it was including the total bilirubin (9.3 mg/dl, which was increased 9 points), direct bilirubin (6.9 mg/dl, which increasing 23 points), SGOT (217 U/L, which increasing 8 times), SGPT (142 U/L, which increasing 4 times), and the Gamma GT (120 U/L, which increasing 3 times). Another laboratory study showed that was slightly increasing of the Prothrombine time (13,1 second). Thyroid function revealed normal. The CMV serology showed that the IgM CMV was negative but the IgG anti CMV was positive. Urine analysis showed there was no sign of urinary tract infection. No microorganism was growth on the urine culture. From two phase of liver ultrasonography we found there was no enlargement of the gallbladder. However another finding showed a thickening of on the anterior portal vein wall, suspected a biliary atresia. With all of those data, the patient was diagnosed as extrahepatal cholestasis et cause suspect biliary atresia.

Patient was proceeded for cholangiography for diagnostic and therapeutic if biliary atresia confirmed intraoperatively. Cholangiography was showed that the contrast has not filled full the gallbladder which its mean confirmation for Biliary Atresia condition. For this reason, the Kasai procedure was performed right away. The liver tissue was taken and sent to laboratory for pathology anatomy examination which then revealed a Chronic Biliary Hepatitis.

Post Kasai procedure, patient was observed and treated in PICU. She received antibiotic combination including Gentamy cin 12.5mg/kgBW every 8 hours combined with Amoxycilin 25 mg/kg every

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Biliary Atresia (BA) is a progressive, fibroobliterative cholangiopathy that affects both the extra- and intrahepatic biliary tree to various degrees, resulting in obstructive bile flow, jaundice in neonates, cholestasis, and cirrhosis. All infants with biliary atresia initially exhibit jaundice. In our cases, all of the three cases came with chief complaint of icteric skin. They were notified to have continuous evaluation, prepared for liver transplantation, and avoidance of any hepatotoxic drugs.

DISCUSSION

Biliary Atresia (BA) is a progressive, fibroobliterative cholangiopathy that affects both the extra- and intrahepatic biliary tree to various degrees, resulting in obstructive bile flow, jaundice in neonates, cholestasis, and cirrhosis. All infants with biliary atresia initially exhibit jaundice. In our cases, all of the three cases came with chief complaint of icteric skin. They were notified to have continuous evaluation, prepared for liver transplantation, and avoidance of any hepatotoxic drugs.

Sequences of para-clinical investigation were proceed following the initial finding in infant with suspicion to BA. One of them is two phase of abdominal ultrasonography which has gained importance as the key diagnostic tool for BA. This non-invasive diagnostic tool is use as the guide before the decision of surgery intervention. This is an operator-dependent tool and can be achieved only in centers where dedicated sinologist are available, it has an overall accuracy of 98%. Further, many previous studies have reported several ultrasound findings to diagnose BA, including a triangular cord sign, abnormal gallbladder length and shape, invisible common bile duct (CBD), and subscapular flow on color Doppler ultrasound. The absence of a gallbladder is as specific as the triangular cord sign in the diagnosis of BA. Other ultrasound characteristic are less valuable findings for diagnosis. A detailed ultrasound examination for the triangular cord sign and for gallbladder abnormalities could reduce the need for liver biopsies and hepatobiliary scintigraphy for infants suspected to have biliary atresia. In our cases, all of the three cases were performed for two phase of abdominal ultrasound. Two of them were came with positive sign of triangular cord as hyperechoic area found on the anterior edge of the portal vein wall, 4 mm in width.

The treatment of BA involves a sequential strategy combining Kasai Portoenterostomy (KPE) as first line and Liver Transplantation (LT) as second line treatment if KPE fails to establish bile flow and/or progressive liver fibrosis and cirrhosis occurs. KPE involves excision of the extrahepatic biliary tree, with reestablishment of bile flow via Roux-en-Y segment of intestine sewn directly to the liver at the portal plate. The outcome of KPE is evaluated by the clearance of jaundice (total bilirubin <2 mg/dL) in 3 months, and the survival of the
children with their own liver. Several factors has been shown affecting the outcome of KPE. The outcome is worse in the embryonic or syndromic form compared to the perinatal form because this form represents an early origin of the disease. Ohio type 3 pattern of atresia, the commonest form of BA, is associated with a poor outcome because in this variant the hilar ductules are also atretic in addition to the extrahepatic biliary system. The most significant factor correlating with success of KPE is the infant's age at the time of surgery.\textsuperscript{13,14}

Among infants who undergo the KPE procedure within 60 days of life, 57%-90% will have successful reestablishment of bile flow, but the success rate drops to <20% if the procedure is performed after 90 days of life.\textsuperscript{8} Despite this worldwide adoption, clear evidence supporting this critical operative time is still lacking.\textsuperscript{27} In our cases, two cases were underwent KPE at the 90 days of age, one other was underwent beyond 90 days of age (it was 4 months old in fact). They were all came with different outcome, one suffered from repeated Cholangitis, one with repeated abdominal leakage, and other without complication. All of this three cases were survived until this case report was made. Regarding to this condition, we search journal based on the evidence and get a journal with entitled of “The anatomic pattern of biliary atresia identified at time of Kasai hepatoportoenterostomy and early postoperative clearance of jaundice are significant predictors of transplant-free survival”, Superina R, Magee JC, Brandt ML, Healey PJ, Tiao G, Ryckman F, Karrier FM, Lyer K, Fectue A, West K, Burns RC, Flake A, Lee H, Lowell JA, Dillon P, Colombani P, Ricketts R, Li Y, Moore J, Wang KS. Ann Surg.2011;254(4):577-585. This journal concluded that age does appear to impact survival with one's native liver. Anatomic pattern of BA, BASM, presence of ascites and nodular liver appearance at KPE, and early postoperative jaundice clearance are significant predictors of transplant-free survival. The risk of transplant/death was significantly lower in the 45.6% of patients who achieved successful bile drainage within 3 months post-KPE (HR 0.08, p<0.001). Then in conclusion they said that survival of BA patients has greatly increased in the era of LT. However many patients surviving with native livers after Kasai operation continue to have signs of biliary cirrhosis and abnormal liver function.\textsuperscript{27} In our three cases, none of them having LT yet. Even though they came with different outcomes, they were all informed about the possibility of liver transplantation one day.

The need for liver transplantation were then more explained on the journal of “Long-term results of biliary atresia in the era of liver transplantation” Lee S, Park H, Moon S, Jung S, Kim J, Kwon C, Kim S, Joh J, Seo J, Lee S.Pediatr Surg Int. 2013;29;1297-1301. On that study, they found that out of 72 patients, 59 received Kasai operation and 13 received LT without prior Kasai operation. Twenty-seven patients received LT after Kasai operation. Survival with native liver was 39% at 10 years. With the application of LT, overall 10-year survival for patients with BA was 94.9%. Among patients alive with native livers after Kasai operation, 14 patients (58.3%) have at least one complication associated with biliary cirrhosis and portal hypertension. Age at which Kasai operation was performed (60days) and postoperative normalization of bilirubin were independent risk factors for survival with the native liver, according to multivariate analysis (HR 2.90, p=0.033, and HR 9.89, p= 0.002). Then in conclusion they said that survival of BA patients has greatly increased in the era of LT. However many patients surviving with native livers after Kasai operation continue to have signs of biliary cirrhosis and abnormal liver function.\textsuperscript{27} In our three cases, none of them having LT yet. Even though they came with different outcomes, they were all informed about the possibility of liver transplantation one day.
The diagnosis can be confirmed by blood culture. The cause of cholangitis is not clear but there is must be an intestinal-biliary communication and therefore the most favored hypothesis is that of an ascending infection from the gut.\textsuperscript{13,27} About this problem, we found journal that this condition can be the the short-term complication of BA post Kasai, \textit{“Surgical outcome and etiologic heterogeneity of infants with biliary atresia who received Kasai operation less than 60 days after birth. A Retrospective study”} Song Z, Dong R, Shen Z, Chen G, Yang Y, Zheng S. Medicine. 2017; 96(26): 1-5. This journal concluded that There were an impact of etiologic heterogeneity on incidence of cholangitis. This study showed that cystic BA had the lowest incidence of cholangitis (15.8%) as compared to the other three BA groups (p=0.011). Next, related to the surgery age, infants with CMV-associated BA who underwent surgery between 51 and 60 days of age exhibited the lowest incidence of cholangitis (33.3%), as compared with infants who underwent surgery at less than 40 days of age (50% incidence) or between 41 and 50 days of age (40% incidence) (p=0.045). A similar trend was observed in infants with syndromic BA and associated malformations. Infants who underwent surgery between 51 dan 60 days of age exhibited the lowest incidence of cholangitis (33.4%), as compared with infants who underwent surgery at less than 40 days of age (50% incidence) or between 41 dan 50 days of age (42.9% incidence) (p=0.027). This result suggest that cystic BA has lowest incidence in BA infants. Both clinical etiologic heterogeneity and operation age may influence BA prognosis.\textsuperscript{24}

Still on discussion of cholangitis after KPE, ascending cholangitis is a frequent and often recurrent complication after surgery.\textsuperscript{23} Recurrent cholangitis causes secondary failure of the restoration of bile flow and possible exacerbation of portal hypertension, resulting in an increase in the risk of mortality and worsened prognosis. Therefore, prevention of cholangitis is important for patients with BA who have undergone KPE.\textsuperscript{23,24,33} Previous studies demonstrated some oral antibiotics such as trimethoprim-sulfamethoxazole and neomycin to be effective as prophylactic agents against ascending cholangitis.\textsuperscript{31} A long-term antibiotic use may, however, increase potential to develop antibiotic resistance and the psychological burden of the patient’s caregiver, suggesting the possibility of decreased compliance to antibiotic treatment.\textsuperscript{23,24} Therefore, other safe prophylactic agents, in addition to antibiotics, for ascending cholangitis are needed. Regarding to this condition, we searched the evidence, we then found the journal of “Use of Lactobacillus casei rhamnosus to prevent Cholangitis in Biliary Atresia after Kasai operation” Lien T, Bu L, Wu J, Chen H, Chen A, Lai M, Shih H, Lee I, Hsu H, Ni Y, and Chang M. JPGN. 2015;60:654-658, which was said that the use of \textit{L casei rhamnosus} was effective as neomycin in preventing cholangitis in patients with BA who underwent Kasai operation and therefore could be considered as potential alternative prophylactic regimen.\textsuperscript{23} In our cases, all of the three patients were having both of antibiotic and probiotic take home after the KPE as prophylactic agent of cholangitis. Among cases, only one of the three cases having recurrent cholangitis after KPE even with both of prophylactic agents.

**SUMMARY**

Three cases of BA were reported in our institution during 2016. They were two cases of two months old female and four months old male. Birth weight were 2100 grams, 3800 grams, and 2770 grams, respectively. All came with persisted jaundice skin > 14 days of life. The laboratory showed elevated direct bilirubin serum, with increasing of GGT up to 22 times. Three portion of stool were acholic. Diagnosis confirmed with Cholangiography. Three of them underwent KPE. After surgery, one suffered from repeated Cholangitis, one with repeated abdominal leakage, and other without complication. The prognosis of BA in children is poor if left untreated. All of these three cases were survived until this case report was made.

**REFERENCES**


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CASE ILLUSTRATION

24. Lien T, Bu L, Wu J, Chen H, Chen A, Lai M, Shih H, Lee I, 
23. D’Agata ID, Balistreri WF. Evaluation of Liver Disease in the 
22. Bezerra JA. Novel approaches to the treatment of Biliary 
21. Ramachandran P, Safwan M, Reddy MS, Rela M. 
20. Kelly DA, Davenport M. Current Management of Biliary 
18. Lee WS, Chai PF. Clinical Features Differentiating Biliary 
17. Shneider BL. Screening for Biliary Atresia: A Ray of Hope. 
15. Superina R, Magee JC, Brandt ML, Healey PJ, Tiao G, 
13. Willot S, Uhlen S, Michaud L, Briand G, Bonnevalle M, 
8. Ultrasound for the Diagnosis of Biliary Atresia: A Meta Analysis. AJR.2016;206;W1-W10 
7. Willot S, Uhlen S, Michaud L, Briand G, Bonnevalle M, 
6. Steir R, Gottrand F. Effect of Ursodeoxycholic Acid on 
5. Liver Function in Children after successful surgery for 
2. Long-Term Outcome of Children with Biliary Atresia after 

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