CASE REPORT

Challenges in Managing Portal Hypertension and Fibrosis in a Case of Biliary Atresia Post-Kasai Procedure—Implications for Early Detection and Long-Term Care

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ABSTRACT

Biliary atresia frequently causes portal hypertension, resulting in significant morbidity and mortality. Elevated portal pressure can be detected as early as during a Kasai procedure. Pediatric portal hypertension is the primary cause of various complications, including variceal hemorrhage, ascites, and extra-hepatic processes. This paper aims to draw attention to the complications and limitations of the Kasai procedure by presenting a case of portal hypertension and fibrosis in an 8-month-old male infant with biliary atresia who underwent the procedure at 3 months. The patient was never completely free of jaundice post-surgery. The jaundice worsened, and the pale-colored stool reappeared two months later. The abdomen was distended, accompanied by ascites, hepatomegaly, splenomegaly, and dilated veins. Edema was present on the lower extremities and the scrotum. The patient was diagnosed with biliary atresia and portal hypertension. Supportive therapy was recommended as a preparatory measure before a liver transplant. However, the parents declined the procedure due to financial constraints. This case suggests that the Kasai procedure may not always be effective. Despite the timely execution of the procedure, liver fibrosis may persist and be associated with portal hypertension. Most patients develop significant fibrosis that progresses to cirrhosis, requiring a liver transplant. To date, biliary atresia remains the primary indication for liver transplant in children, with no alternative medical treatment recognized. This case report highlights the progression of portal hypertension and liver fibrosis following the Kasai procedure for biliary atresia, emphasizing the challenges in early detection, complications management, and the exploration of alternative therapeutic strategies.

Keywords: Biliary atresia; portal hypertension; Kasai procedure; child mortality

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Highlights:

1. This study critically reviews the limitations and complications of the Kasai procedure in treating biliary atresia, with particular focus on portal hypertension issues, for which case reports are limited.
2. This study highlights the important need for accessible and effective long-term treatment alternatives by presenting a pediatric case where financial constraints prevented a liver transplant.
3. The data from this study are anticipated to contribute to the advancement of healthcare equity and pediatric liver disease management.

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INTRODUCTION

Biliary atresia is a condition that specifically impacts the extrahepatic and intrahepatic bile ducts of the liver, resulting in cholangiopathy. In Asia, particularly Taiwan and Japan, the incidence rate is significantly high, ranging from 100 to 500 cases per 100,000 live births. In contrast, Europe exhibits an incidence rate between 5 and 25 cases per 100,000 live births. The current therapy options for this condition are restricted to a surgical technique known as the Kasai portoenterostomy. However, the Kasai portoenterostomy is ineffective for improving the condition in over half of the patients, leading to unresolved intrahepatic cholangiopathy. Moreover, biliary atresia often results in the development of fibrosis, portal hypertension, and liver failure. Patients with biliary atresia frequently require liver transplant, which in turn necessitates lifelong immunosuppression. This intervention has a significant impact on the quality of life of these patients (Lendahl et al., 2021).

Portal hypertension arises from an increase in the resistance of blood vessels within the portal circulation, an elevation in blood flow in the portal vein, or a combination of both factors. Pulmonary hypertension is typically diagnosed by identifying related problems, such as multiple portosystemic shunts, ascites, and hepatic encephalopathy. The management of portal hypertension primarily aims to

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control these problems (Buob et al., 2011).

Portal hypertension is a frequent result of biliary atresia and can have serious consequences for patients, which may lead to significant morbidity and mortality. The onset of portal pressure elevation can be observed as early as during a hepatoportoenterostomy. Portal hypertension in children can lead to variety of complications, including variceal hemorrhage and ascites. Additionally, it can also cause extra-hepatic processes, such as hepatic pulmonary syndrome, hepatic renal syndrome, porto-pulmonary hypertension, and hepatic encephalopathy (Chiou & Abdel-Hady, 2017).

This case report aims to highlight the complications and limitations associated with the Kasai procedure in the treatment of biliary atresia, particularly focusing on the progression to portal hypertension and subsequent need for liver transplant. This report provides an overview of the clinical course and challenges faced by a pediatric patient with biliary atresia after the Kasai procedure, emphasizing the persistent problems of jaundice, liver fibrosis, and portal hypertension despite timely surgical intervention. Additionally, this report underscores the critical need for liver transplant as the definitive treatment for biliary atresia, given the significant morbidity and mortality associated with portal hypertension.

CASE REPORT

An 8-month-old male infant presented to the emergency department with complaints of jaundice, distended abdomen, and swollen hands and feet. Over the previous two days, the patient exhibited black stool, frequent itching, skin redness, and swelling in the right scrotum. The medical history indicated that at 3 months of age, the patient had presented to the Pediatric Hepatology Clinic at Dr. Soetomo General Academic Hospital, Surabaya, Indonesia, with a chief complaint of prolonged jaundice lasting three days. The patient's stool was pale, and the urine was dark yellow. The patient exhibited neither fever nor bleeding. After receiving a diagnosis of biliary atresia, the patient subsequently underwent the Kasai procedure. The patient was in a stable condition following the surgery. One week after the surgery, the patient's stool started to appear yellow, as compared to its previous pale coloration. The patient was never entirely jaundice-free after the surgery. However, the parents reported that the jaundice had been alleviated. At the age of 4 months, the patient's jaundice persisted, although pale stool was denied. At the age of 5 months, the patient presented with a fever that had lasted for five days. The parents reported that the jaundice had exacerbated. In addition, the patient's abdomen enlarged, and the pale-colored stool reappeared.

The patient was born via cesarean section at a gestational age of 38 weeks with fetal distress. The infant weighed 3,320 g and measured 48 cm in length. The baby's first cry occurred immediately after the delivery. The baby was breastfed from birth to two months old and gained adequate weight. There was no family history of a similar disease. The physical examination revealed icteric sclera, as shown in Figure 1. The abdominal examination indicated a distended abdomen with ascites, an enlarged liver, an enlarged spleen, and dilated veins, as displayed in Figure 2. Edema was present on both the lower extremities and the scrotum.

The upper arm circumference measured during an anthropometric examination indicated severe malnutrition. The laboratory examination confirmed that the patient had anemia, indicated by a hemoglobin level of 9.7 g/dL, as well as leukocytosis, determined by a white blood cell count of 28,900 x 103/µL. The liver function biomarkers showed a direct bilirubin level of 12.85 mg/dL, a total bilirubin level of 19.38 mg/dL, and an albumin level of 2.35 g/dL. Furthermore, the biomarker examination yielded the following results: an aspartate aminotransferase (AST) level of 139 u/L, an alanine aminotransferase (ALT) level of 191 u/L, a prothrombin time (PT) of 20 seconds, and an activated partial thromboplastin time (APTT) time of 47.9 seconds. The serum electrolytes were within the normal range. A preoperative Doppler abdominal ultrasound revealed mild hepatomegaly and gallbladder hypoplasia. The color Doppler flow imaging of the hepatic, portal, and renal veins was normal. After the surgery, the Doppler abdominal ultrasound revealed parenchymal liver disease, an increased portal vein flow velocity, and splenomegaly (Figure 3). A liver biopsy indicated extrahepatic cholestasis accompanied by bridging fibrosis, also known as stage 3 fibrosis (F3), which was consistent with biliary atresia.

According to the examination results, the patient was diagnosed with biliary atresia and portal hypertension. Propanolol, furosemide, spironolactone, ursodeoxycholic acid, albumin transfusion, and fresh frozen plasma (FFP) transfusion were administered as supportive therapy to the patient. Initially, the patient was prepared for a liver transplant. Despite the recommendation, the parents declined the procedure owing to financial constraints.

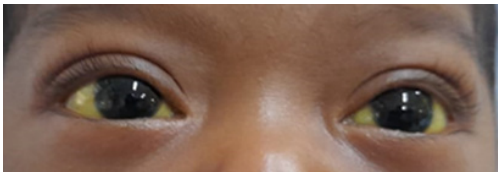

Figure 1. Clinical manifestations of icteric sclera in the patient

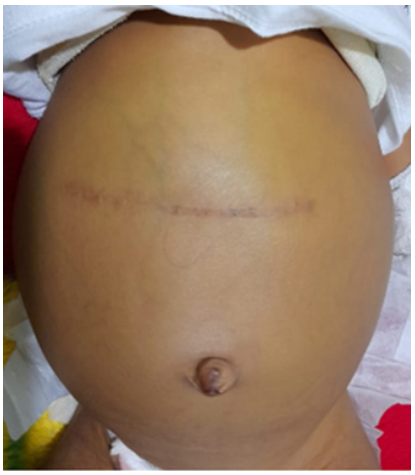

Figure 2. Physical examination shows abdominal enlargement, hepatomegaly, splenomegaly, and venectasis of the abdominal veins

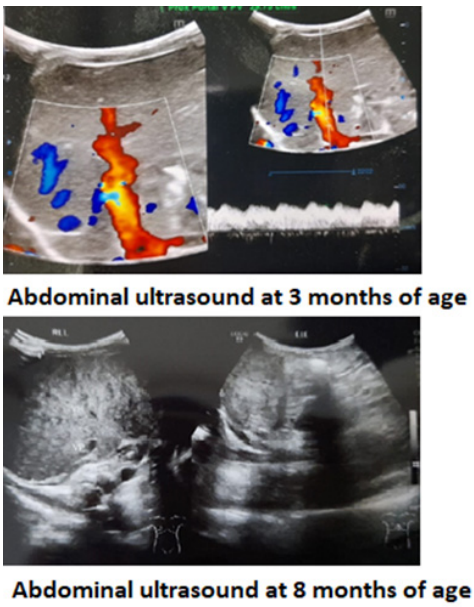

Figure 3. Abdominal ultrasound evaluation of the patient

DISCUSSION

Biliary atresia is a progressive, idiopathic, and fibro-obliterative condition that affects the extrahepatic biliary tree. This condition is characterized by biliary obstruction that occurs solely during the neonatal period. The injury may result in scarring, a reduction in liver tissue and function, and the development of cirrhosis (Bezerra et al., 2018). Biliary atresia signifies the primary indication for liver transplant in pediatric patients. To date, no alternative medical treatments have been recognized as effective (Wang et al., 2015).

The clinical manifestations of biliary atresia typically become apparent within one to six weeks following birth. The patients experience a gradual worsening of cholestasis, resulting from the liver's failure to excrete bile. When the liver fails to excrete bilirubin via the bile ducts, bilirubin starts to build up in the bloodstream, leading to various symptoms. The symptoms observed include a yellowing of the skin, itchiness, inadequate nutrient absorption leading to delayed growth, pale stools, darkened urine, and abdominal distension. Ultimately, the progression will lead to the development of cirrhosis accompanied by portal hypertension. Untreated biliary atresia may result in liver failure (Couturier et al., 2015). In this case, the pediatric patient presented with a chief complaint of prolonged jaundice. The complaint was also accompanied by pale-colored stools and dark urine, suspected as cholestasis leading to biliary atresia.

Biliary atresia results in impaired bile flow from the liver, causing bile to accumulate within the liver cells, a condition known as cholestasis. This accumulation leads to inflammation and fibrosis, resulting in the gradual scarring of liver tissue. As the condition advances, it leads to increased stiffness and constriction of the liver's blood vessels, especially the small veins within the liver, known as sinusoids. This elevates the obstruction to blood circulation within the liver. The portal vein transports blood from the intestines and other organs to the liver for detoxification and nutrient processing. However, it encounters heightened pressure while attempting to move blood through the fibrotic and resistant liver. This results in increased pressure within the portal vein system, which is referred to as portal hypertension (Morales-Ruiz et al., 2015). Portal hypertension is a primary factor contributing to illness and mortality in pediatric patients with liver disease (Alukal & Thuluvath, 2019).

Portal hypertension is a condition that arises when there is either an elevation in blood flow or an increase in resistance inside the portal system. Portal hypertension is categorized according to the specific anatomical site where the condition hinders the blood flow in the portal system: it may occur before the liver (prehepatic), within the liver (intrahepatic), or after the liver (posthepatic). Portal hypertension is defined as an increase in portal pressure of 10–12 mmHg or a hepatic venous pressure gradient exceeding 4 mmHg. The typical range for portal pressure is 5 to 10 mmHg (Oliver et al., 2024). In this case, the patient was diagnosed with biliary atresia after the Kasai procedure and subsequently developed portal hypertension as a complication, indicated by the presence of bloody stools, frequent itching, reddish skin, and swelling of the right scrotum. The portal hypertension likely developed as a consequence of biliary cirrhosis affecting the liver's blood flow. We aimed to highlight the challenges associated with the Kasai procedure, particularly the risk of portal hypertension and liver fibrosis, emphasizing the need for early detection and proactive management strategies to improve outcomes for affected children.

The clinical signs and physical examination findings of portal hypertension vary according to the root cause and the extent of hepatic dysfunction and fibrosis. The manifestations of portal hypertension encompass hemorrhaging, splenomegaly, and fluid accumulation in the abdominal cavity known as ascites (Yayla et al., 2023). Common systemic symptoms in children include fatigue, muscle weakness, loss of appetite, nausea, vomiting, and failure to thrive (Bosch & Iwakiri, 2018).

Patients exhibiting symptoms of portal hypertension require an assessment to identify the underlying cause of liver malfunction, determine the extent of fibrosis, and evaluate the existence and severity of any consequences outside the liver. Other diagnostic tests may also be taken into consideration. Furthermore, it is recommended that all patients undergo an abdominal ultrasonography with Doppler to assess the liver's echotexture and rule out any structural or vascular irregularities (El-Nakeep & Ziska, 2024).

The most serious complication of portal hypertension is gastrointestinal bleeding, which is usually caused by portal vein thrombosis. Children develop conditions that result in gastrointestinal bleeding during their first decade of life (Alukal & Thuluvath, 2019). Various complications can arise from portal hypertension, such as enlargement of the spleen, the formation of collateral blood vessels, the development of varicose veins in the gastrointestinal tract, damage to the stomach lining, problems in the colon and intestines, low levels of either red and white blood cells, platelets, or all blood cells, increased blood flow, the accumulation of fluid in the abdomen, kidney problems, bacterial infections, impaired brain function, and lung and heart conditions (Pedersen et al., 2015; Oliver et al., 2024).

Malnutrition frequently occurs in individuals with cirrhosis as a result of portal hypertension, which leads to reduced appetite and insufficient food consumption. Additionally, the condition is exacerbated by increased metabolic requirements, impaired absorption of nutrients, excessive fat in the stool, and deficiencies in fat-soluble vitamins. Chronic malnutrition leads to the development of stunted growth in children with cirrhosis, which is linked

to unfavorable outcomes (Banc-Husu et al., 2023).

In the case reported in this study, ensuring the prevention of severe consequences is crucial in the therapy of the patient's portal hypertension. A non-selective beta blocker was administered as part of the medical treatment. Propanolol, a beta blocker that does not have a specific target, is efficacious in treating pediatric conditions that result in portal hypertension (Garcia-Tsao et al., 2017). Spironolactone, an aldosterone antagonist, is the preferred choice for diuretic treatment in individuals with liver disease (Oliver et al., 2024). Aldosterone antagonists counteract the excessive production of aldosterone, which is a characteristic of ascites resulting from portal hypertension. They specifically oppose the salt retention induced by aldosterone, leading to a mild increase in urine production. In addition, albumin infusions were administered. The administration of albumin can elevate intravascular osmotic pressure, thereby facilitating the efficacy of diuretics to stimulate urination (Campos-Munoz et al., 2024).

Cirrhosis causes not only decreased albumin synthesis but also specific structural and functional changes, such as post-transcriptional alterations and oxidative damage, which have been linked to impaired function and unfavorable clinical outcomes. The patient in this case report was administered ursodeoxycholic acid. The medication is a hydrophilic bile acid that has become more commonly used for the treatment of several cholestatic diseases. Nevertheless, the monitoring of drug-related side effects and complications from portal hypertension is still ongoing. This is because portal hypertension in biliary atresia is usually progressive. As the patient's prognosis was poor, the only viable treatment option was liver transplantation (Ge et al., 2020). This patient urgently required a liver transplant due to the rapid progression of the disease. Unfortunately, liver transplantation could not be performed due to financial limitations and inadequate facilities.

The only method for accurately and reliably diagnosing and treating biliary atresia is through surgical exploration. The Kasai procedure is a recognized surgical intervention aimed at reestablishing bile drainage in individuals affected by biliary atresia (Chen et al., 2022). Timely surgical intervention is essential for the preservation of native liver function and the overall survival of the patient. The standard Kasai procedure involves creating an enterostomy approximately 10 cm distal to the ligament of Treitz, facilitating the formation of a double-Y hepatic portojejunostomy. The distal cut is elevated to the porta hepatis to facilitate an anastomosis, with the cut surface of the fibrous mass located at the porta hepatis. The proximal cut is subsequently positioned adjacent to the jejunum, approximately 45 cm distal to the ligament of Treitz, where an end-to-side anastomosis is performed to establish the Roux-en-Y configuration (de Carvalho et al., 2019).

The disadvantage of the Kasai procedure is that it may not always be effective. Even if the Kasai procedure is carried out on time, liver fibrosis may persist. Portal hypertension associated with liver fibrosis may also arise, as reported in a substantial number of patients. The success rate for achieving optimal bile flow following the Kasai procedure varies depending on the age of the patient. However, it notably increases to 90% when the procedure is conducted before the age of 8 weeks. The Kasai procedure offers certain immediate benefits, including effective decompression and drainage that safeguard against cirrhosis and promote healthy growth until a successful liver transplant is achieved (Qiao et al., 2015; Guérin et al., 2019). Unfortunately, even after the Kasai procedure is performed, the majority of patients develop significant fibrosis that progresses to cirrhosis, requiring liver transplantation (Pedersen et al., 2015; Xu et al., 2023).

The progression of liver fibrosis remains a critical issue in patients following the post-Kasai procedure, even in those with initially successful bile drainage. While the Kasai procedure aims to delay the need for liver transplantation, it does not prevent the continued scarring of the liver tissue. Around 60% of infants with biliary atresia initially experience restored bile flow following the Kasai portoenterostomy. However, hepatic fibrosis tends to progress, and portal hypertension develops in most children who undergo the procedure (Sundaram et al., 2017). In this case, biliary cirrhosis likely contributed to the patient's portal hypertension, a scenario that underscores the importance of early identification and monitoring of fibrosis.

Non-invasive methods (e.g., elastography) and serum biomarkers (e.g., matrix metalloproteinase-7) may provide insights into the progression of fibrosis. However, therapeutic options to reverse or slow down fibrosis in pediatric populations are currently limited. Early transplantation referral, particularly when signs of decompensation emerge, remains the cornerstone of managing advanced liver disease in biliary atresia patients (Jiang et al., 2024).

Early detection of biliary atresia through screening programs has proven effective in improving surgical outcomes. For example, stool color cards have shown the ability to improve the timeliness of diagnosis, facilitating earlier Kasai procedures. However, this case demonstrated that despite early intervention, the risk of progressive fibrosis and its complications persists, necessitating lifelong surveillance (Gu et al., 2015; Madadi-Sanjani et al., 2021).

This case report elucidates the complications and constraints associated with the Kasai procedure for treating biliary atresia. The pediatric case demonstrated that, despite the timely procedure, there may potentially be persistent problems, such as jaundice, liver fibrosis, and portal hypertension. This highlights the urgent need for liver transplant as the cornerstone of biliary atresia treatment. However, this case report has several limitations. Firstly, the study only included a single case, which may limit the generalizability of the findings to a wider population of children with biliary atresia. Secondly, due to time constraints, the study did not provide long-term follow-up data, which could offer a more comprehensive insight into the outcome and effectiveness of the Kasai procedure and subsequent treatment.

Long-term care for patients post-Kasai procedure should focus on regular monitoring for complications, such as portal hypertension and cirrhosis. Multidisciplinary teams comprising hepatologists, gastroenterologists, and surgeons should coordinate in the care of these patients. In the absence of liver transplant, it is vital to focus on symptom management, nutritional assistance, and surveillance for variceal bleeding to improve patient outcomes.

SUMMARY

Early diagnosis of biliary atresia is crucial for improving patient outcomes. Infants presenting with new-onset jaundice or jaundice persisting beyond two weeks should undergo testing for total and conjugated bilirubin levels to rule out cholestasis. The use of a stool color card is effective for parents to help identify potential biliary atresia. If left untreated, biliary atresia can lead to rapid progression of liver fibrosis, portal hypertension, and end-stage liver disease, often resulting in mortality within the first two years of life. This case underscores the need for heightened awareness and prompt intervention to mitigate these serious complications and emphasizes the importance of ongoing research and education regarding early detection strategies.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

PATIENT CONSENT FOR PUBLICATION

This case report has been approved for publication by the patient's parents acting as the legal guardians.

FUNDING DISCLOSURE

None,

AUTHOR CONTRIBUTION

All authors contributed to every phase of this case report, including interpretation of the data, drafting of the article, critical revision of the article for important intellectual content, final approval of the article, provision of administrative, technical, or logistic support, and collection and assembly of the data.

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