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Review Article

Summarization of the literature published on biliary atresia: a comprehensive literature review

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ABSTRACT

The leading cause of pediatric liver failure and liver transplantation is biliary atresia. Early diagnosis of BA is challenging, and delayed diagnosis caused many complications. Most BA patients eventually went through liver transplantation even if received the initial treatment of the disease. The incidence of BA is rare hence limited literature has been published about diagnosis, treatment, risk factors and survival rate among BA patients. Therefore, this literature review aimed to provide an opportunity for physicians and researchers to know about different diagnostic and treatment tools as well as about the risk factors causing variation in the survival rate after liver transplantation. Therefore, knowledge about the advanced methods of diagnosis provides an early and accurate diagnosis and providing treatment with fewer risk factors help to increase favorable outcome and increase the survival rate among BA patients.

Keywords: Biliary atresia, Pediatric, Diagnosis, Treatment, Risk factors, Survival rate

INTRODUCTION

The progressive cholangiopathy with fibro-obliterative obstruction of the biliary tract is known as biliary atresia (BA).[1] It affects both intrahepatic and extrahepatic tracts which cause cirrhosis, liver failure and sometimes death if not timely and properly treated.[2] The widely accepted hypothesized cause of biliary duct injury is initially an infection followed by an autoimmune response caused by the infection, leading to damage to the biliary tract.[3]

The clear etiology of BA still remained unknown despite its delayed diagnosis and poor prognosis. This disease is quite similar to other medical conditions like physiological neonatal icteric, hence initial findings are usually quite confusing and thus diagnosis of BA is delayed.[4] However, timely diagnosis of BA and accurate differentiation it from other medical conditions is the key to survival from the disease. However, the initial treatment course among 70-80% of the children with BA is portoenterostomy and eventually, liver transplantation is required after the procedure.[5] According to the literature, the incidence of biliary atresia is more common is Asia (1:8000) compared to Europe or America (1:15000-20000).[6,7] Chiang et al reported that the incidence of BA among Asia-pacific countries is as high as 1:3300 as compared to Europe at 1:18000.[8] Risk factors associated with BA incidence have been studied by researchers and reported. Genetical factors or chromosome alteration, viral-induced immune dysregulation, and maternal disease are common risk factors that cause the incidence of BA.[7] The study by Fischler et al suggested maternal and gestational age, parity and gravidity, intrauterine growth retardation (IUGR), birth weight and race/ethnicity as pediatric BA risk factors.[9] Limited studies reported the incidence of BA around the globe. Therefore, very limited literature is available reporting the etiology, diagnostic methods, treatment methods and risk factors associated with BA.

Therefore, the scientific and medical community perhaps has limited knowledge about the various diagnostic tools and treatment strategies used among the institutions and hospitals situated across the globe. Hence, this provided the rationale to conduct a literature review of the studies published on the topic of BA and provide a summary of the work published so far.

The present literature review also reported the demographics of the BA patients in the reported studies, methods used for diagnosis and their accuracy, treatment strategies and their success rate, potential risk factors and short- and long-term survival rate of BA patients after treatment.

METHODS

A literature search was conducted in January 2023 in which studies published under the topic of BA were searched. All types of original articles, reviews or case reports were searched and included in the present literature review.

The literature was searched on the following databases; Scopus, Web of Science (WOS), Pubmed and Summon. The keywords used for the literature search were 'pediatric biliary atresia'. The keywords were searched in the title, abstract and article keywords, and all those in which any search keyword was found were extracted. As a result, 6 publications were found at Pubmed, 7 were at WOS, 10 were at Scopus and 12 were at Summon. Hence, a total of 35 publications were found. Titles of all first searched outcomes were reviewed to remove the duplication. A total of 21 duplications were found and removed and there were 14 titles remaining. The second phase of the literature search consisted of retrieving the of full text of those 14 publications. As an outcome, 11 full-text publications were found and included in the present literature review. After retrieving the full text, the contents of the studies were screened. It was found that the studies could be divided into the following categories: demographics of the patients included in the studies, type of diagnostic methods used, and different kinds of treatment methods used to treat BA patients, potential risk factors and long-and-short term survival rate of the patients after having LT.

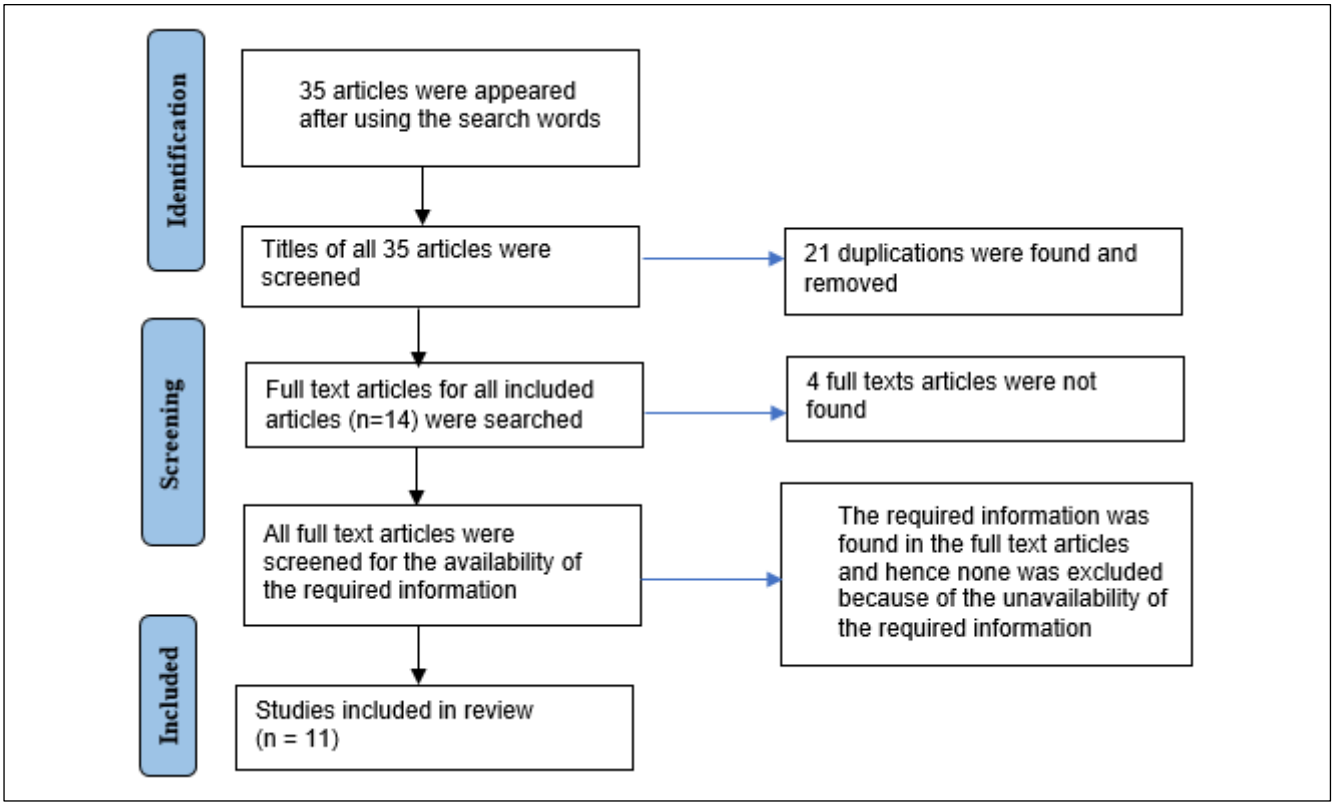

Figure 1: Identification of studies from the databases

DEMOGRAPHICS OF THE PUBLISHED STUDIES

Among the 11 published articles included in the present review, 9 (81.8%) were original articles, 1 (9.1%) review article and 1 (9.1%) case report were found. Regarding the year of publication, 4 (36.4%) out of 11 articles were published in 2019 which was the highest number with respect to the year of publication (Figure 2).

The sum of the number of patients reported in the articles included in the present review was 2621. The highest number of BA patients was included in the study published by Kasahara et al in 2018.[10] The study used the data repository to extract all the BA patients' data reported between 1989 and 2015. Hence, due to the long period, total number of extracted cases was 2085. Therefore, around 80% of the total number of cases were found in a

single study. Another study with a relatively large sample size was published by Lin et al in 2020.[11] The study included 291 BA patients however duration considered for data extraction was between 1994 and 2017. Hence, both studies took more than 20 years of data therefore a large number of BA cases were reported. On the other hand, the number of cases reported in other studies was found to be less than 100 even though some studies had less than 10 reported cases.

Analysis for the publication country was also conducted and it was found that 9 (81.8%) out of 11 studies were published from East Asian countries. In which, 4 (44.5%) were published from Japan, 2 (22.2%) from Taiwan, 2 (22.2%) from China and 1 (11.1%) from Korea. In addition, one study was published from Belgium and one from Indonesia.

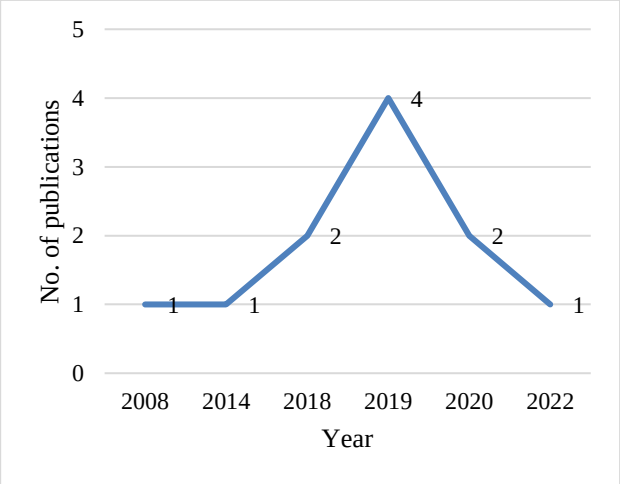

Figure 2: Number of publications per year.

DIAGNOSTIC METHODS

Globally, pediatric liver failure and liver transplantation could be largely due to BA.[12-14] Therefore, early and accurate diagnosis of the disease is a great need. In the past few years, the use of molecular levels as a biomarker caught the most attention and studies have been done on this.[12] Studies reported the high concentration of MMP-7 among BA patients and perhaps serves as a diagnostic parameter.[15,16] BA could be due to immune-related genes (IRGs) and can serve as a biomarker.[17] Li et al conducted a study in which they studied the role of IRGs in the occurrence of BA. The authors used the NCBI database to download the gene expression data. The data consisted of 7 healthy controls and 171 infants with BA. From the analysis, 650 differentially expressed genes (DEGs) were found among the BA and normal in which 146 and 504 genes were downregulated and upregulated respectively. Li et al identified VCAM-1 (vascular cell adhesion molecule-1) as a potential immune biomarker of BA.[17] The authors found a significant variation in VCAM-1 level among the BA patients compared to the control group. In addition, HLA-DRA (human leukocyte antigen DR alpha chain) expression was found in the BA liver and it might play a role in BA. Li et al also reported a significant association between CD74 (MHC class II invariant chain) and the occurrence of BA. In a nutshell, the study identified VCAM-1, HLA-DRA and CD74 as biomarkers of BA.[17]

Due to the complexity of the differentiation of BA and non-BA patients. Tsuda et al in 2019 published a study in which they used ^{99m}Tc-PMT scintigraphy as a diagnostic method of BA.[18] Along with the ^{99m}Tc-PMT scintigraphy, the study used total bilirubin, stool color change, direct bilirubin, aspartate aminotransferase (AST) and γ-glutamyl transferase (γ-GTP) to identify BA patients. It was found that the ^{99m}Tc-PMT scintigraphy had the highest diagnostic accuracy (94.2%) (REFB). Hence, the authors concluded that highly accurate results could be obtained from ^{99m}Tc-PMT scintigraphy compared to the conventional methods.

TREATMENT METHODS

Biliary secretion of lipids, toxic metabolites and xenobiotics is done by the bile acids. These acids are secreted into the bile in the duodenum, reabsorbed at the terminal ileum and through the portal vessels transported back to the liver.[19] Any obstruction in the bile ducts causes many problems. One of the most recognized causes of this obstruction is an injury to the biliary duct which perhaps initially be caused by the infection and the autoimmune response induced by the infection leads to progressive damage to the biliary duct.[3]

Progressive cholangiopathy with fibro-obliteration obstruction of the bile duct is known as BA. It is difficult to diagnose BA at its early stages due to the delayed presentation of its clinical features. Therefore, in many countries, it diagnosis after 60 days of birth.[20,21] Morio Kasai 1959 introduced a BA management technique known as Kasai portoenterostomy (KPE). After three months of KPE, if serum bilirubin<2 mg/dl and jaundice clearance are found in BA patients that indicate the transplant-free survival of the patient.[22]

On the other hand, standard open portoenterostomy (OPE) is another effective way to manage BA patients. Literature reported 47-65% of BA patients after OPE achieved jaundice clearance.[23-26] That is why, in many countries, OPE is the first line of treatment of BA patients.[22]

The first surgical technique for the management of BA was initially used by Esteves et al in 2002.[27] The procedure is named laparoscopic portoenterostomy (LPE), and some studies indicated that the procedure was found to be safe and feasible.[28,29] However, some other studies claimed that an inferior outcome was achieved among the patients who underwent LPE compared to patients who underwent OPE.[30,31] However, after the recent advancement of the technology and instrumentation, some researchers reported the good outcome of the LPE.[32,33] In addition,

there are many advantages of LPE including less tissue injury, no mobilization of the liver, smaller wounds, decreased pain and fewer abdominal adhesions.[28]

In the meta-analysis published by Li et al in 2019, the outcome of the two techniques (OPE and LPE) for the management of BA patients was compared. Results provided that the only operative time was found significantly less for patients underwent OPE compared to patients who underwent LPE. However, other parameters like intraoperative blood loss, cholangitis, early clearance of jaundice or two years of survival with native liver were not found significantly different in the two techniques.[34]

Although the first line of treatments for the management of BA patients is utilized initially. However, the majority of patients will eventually receive liver transplantation.[34] Transplantation of the liver is a critical treatment procedure for patients with liver failure. Living-donor liver transplantation (LDLT) is the most widely used liver transplantation treatment method to save the lives of children with end-stage liver disease.[35] This method was introduced in Japan in 1989 to save the lives of BA patients because of the scarcity of donors available for deceased donor liver transplantation (DDLT).[36]

RISK FACTORS

Research has been conducted to describe the risk factors associated with the incidence of BA as well as the risk factors cause the unsuccessful outcome of the treatment of BA. Risk factors associated with the incidence of BA reported in the literature are genetic factors or chromosome alteration, viral-induced immune dysregulation, maternal disease including gestational diabetes and perhaps some other factors which could potentially be associated with the incidence of BA.[7] In addition, maternal age, parity and gravidity, gestational age, IUGR, birth weight, and race/ethnicity are also reported as risk factors for pediatric BA.[9] A study from Sweden found that mothers more than 35 years old had a higher risk of having children with BA.[9] pregnancy in older maternal age is a fetomaternal factor which is a potential risk factor that causes BA incidence.[37] Diabetic mellitus and hypertension are also reported as risk factors for the incidence of biliary atresia. The vascular dysfunction triggers the hypoxia induced ischemia in fetal liver vascularization.[38]

On the other hand, studies have also been conducted to study the risk factors associated with the treatment outcome of BA. Perkins in 2006 studied the risk factors associated with graft functioning and receivers' survival rate were also studied and reported. It was reported that obesity is a strong risk factor associated with graft function and patients' survival rate.[39] In addition, Kasahara et al reported that the significantly associated risk factors were the donor's body mass index (BMI), graft type, recipient age and center experience.[10] Noguchi et al in 2019 studied acute cellular rejection (ACR) among patients who underwent LDLTs with maternal or paternal grafts. The study included 46 of which 28 had BA, all patients underwent LDLT either with maternal or paternal grafts.

In patients with BA, there was no significant difference in the proportion of rejection between patients who received grafts either from the maternal or paternal side.[35] However, study findings provided that the first rejection appeared significantly earlier among the patients who had grafts from the maternal side compared to the other group. In addition, the analysis also provided that the gender (male) of the donor had a significant association with the first rejection.[35] Contrarily, the study published by Nijagal et al found that the rate of maternal graft failure was lower than paternal graft failure (3.7% verses 10.5%) and therefore required fewer episodes of re-transplantation (2.7% verseLPEs 7.5%).[40]

SURVIVAL RATE OF BA AFTER LIVER TRANSPLANT

Cholesteric liver disease, metabolic liver disease, acute liver failure, hepatic malignancy etc. are the conditions of pediatric liver transplant (LT). BA is another and most common LT condition, accounting for almost 50% of all pediatric LT.[41] In Japan in 1989, a new method of LT was introduced named living donor liver transplantation (LDLT). This method was introduced due to the scarcity of donors and serves as a life-saving procedure, especially for patients with BA.[35]

Many researchers have studied the long-term outcomes of LT in BA patients and reported 5 years survival rate ranging between 82% to 98%.[42-44] However, limited data on LDLT was studied and published. Hence, very limited research is available reporting survival rates among the patients treated with LDLT.

Kasahara et al published an article from Japan in 2017 in which they gathered BA patients' statistics who underwent LDLT between 1989 to 2015. The authors found 2085 BA patients who underwent LDLT and their survival rate was studied. The survival rate was found to decrease due to increased time and the presence of some risk factors. The 1-, 5-, 10- and 20-year graft survival rates were 90.5%, 90.4%, 84.6%, 82.0% and 79.9% respectively.[10]

The graft survival rate among the BA patients who underwent LDLT was also studied by other researchers. Diem et al from Belgium 2003 studied 328 BA patients among which 18% went through LDLT. The authors found 1-, 5- and 10- years survival rates of 77%, 72% and 71% respectively.[45] Similarly, a study published in the USA studied 1976 BA patients among which 16.5% underwent LDLT. The 1-, 5- and 10- years survival rates were reported to be 83.6%, 76.2% and 72.7% respectively.[46] Fouquet et al from France 2005 studied 280 BA patients among which 3% underwent LDLT and 1-, 5- and 10-years graft survival rates were 77%, 73% and 71% respectively.[47] A study from Taiwan had 100 BA patients

and all of them went through LDLT. The 1-, 5- and 10-years graft survival rates were found to be 98%, 98% and 90% respectively.[41]

DISCUSSION

The incidence of pediatric biliary atresia is not very common and is witnessed very rarely. However, its incidence is relatively high among Asia-pacific countries compared to European or American countries.[48] Due to the rare incidence of BA, limited literature available reported BA cases. Hence, information about its diagnosis, treatment, risk factors and survival rate after the treatment is very limited. Therefore, the present literature review was designed to review and gather information published in the articles related to diagnosis, treatment, risk factors and survival rate among BA patients. Due to the relatively high incidence of BA cases in Asia-pacific countries, most of the articles found for the present review were from Asian countries. In addition, most of the literature found for this review was published during the past five years.

Diagnosing BA clinically may be difficult because distinguishing the cause of persistent neonatal jaundice is due to BA or due to some other reasons is difficult.[18] However, early diagnosis of BA is the key to survival.[49,50] On the other hand, clinical parameters and biochemical tests are usually inconclusive.[18] Hence, researchers recommended using of molecular levels as a biomarker for the diagnosis of BA. Studies reported the high concentration of MMP-7, significant variation in VCAM-1, HLA-DRA and CD74 as a biomarker for the diagnosis of BA.[15-17] Tsuda et al suggested using ^{99m}Tc-PMT scintigraphy as a diagnostic method of BA to differentiate BA and non-BA patients. Achieving high diagnostic accuracy the authors revealed the ^{99m}Tc-PMT scintigraphy can produce better results compared to conventional methods.[18]

The first line of treatment method reported in the literature to manage BA cases is OPE. Literature reported that the achievement of jaundice clearance in BA patients was over 60% after standard OPE.[23-26] A laparoscopic method was introduced in 2002 called as LPE. The LPE compared to OPE is relatively a new method for the management of BA cases. Therefore, this method has not been used widely and hence very limited data published on this method. Some studies found this better in terms of less tissue injury, no mobilization of the liver, small wounds etc.[32,33] On the other hand, some studies discourage the use of LPE as a method of BA management.[30,31]

It was found that the risk factors played a significant role to attain a successful outcome in the treatment of BA. As reported in the literature, most BA patients ultimately need to have liver transplantation either due to the unsuccessful outcome of first-line treatment methods or the reoccurrence of the disease.[51] Various risk factors have been reported to have significant associations with the successful outcome of LT and patients' survival. Among the risk factors, the donor's BMI was identified as one of the significant risk factors associated with poor graft function and reported by many studies.[39] In addition to this, gender of the graft donor and ABO incompatibility are also identified as potential factors that affected significantly the successful outcome.[10] Matching of the liver graft size is another factor associated with a successful outcome. Lower graft survival was found in the case of using small-for-size grafts due to their insufficient metabolic and synthetic function.[52]

Similarly, researchers also studied the survival rate among BA patients who underwent liver transplantation. LDLT is a widely used method for the treatment of BA patients. Studies reported 1 to 10 years of survival rate for the patients who underwent LDLT.

The reported one-year survival rate by the researchers was varying between 77% to 98%, 5-year survival was varying between 72% and 98% and 10-year survival was between 71% and 90%.[10,41,45-47] The presence of various risk factors causes variation in the reported survival rates among different studies. Reducing the risk factors perhaps provide higher chances to attain favorable outcome. In addition, it was also found that graft survival was worst among adult BA patients compared to pediatric BA patients.[10]

CONCLUSION

This literature review provided an opportunity for the researchers and physicians to know about the diagnostic and treatment methods, risk factors and survival rate among BA patients. The new techniques for diagnosis provide a chance to early diagnose the disease. Advancements in treatment methods help to treat less pain and less duration of stay at the hospital. Identification of the risk factors contributes to reducing the chances of graft failure as well as increasing the survival rate of the patient. In conclusion, this work contributes to an increased understanding of BA and effective ways to increase successful treatment outcomes.

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