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Japanese biliary atresia registry

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ABSTRACT

The Japanese Biliary Atresia Registry (JBAR) was launched in 1989 by the Japanese Biliary Atresia Society (JBAS) to investigate the epidemiology and etiology of biliary atresia and to improve surgical outcomes. The JBAR collects data through initial, liver transplantation, and follow-up questionnaires. Pediatric surgeons from JBAS member institutions and hospitals affiliated with the Japanese Society of Pediatric Surgeons are responsible for registering patients and submitting data through an online system. Each patient is to be followed up for 40 years. As of 2023, 3951 patients had been registered, with 1688 undergoing liver transplantation. The native liver survival rates in the 10th, 20th, and 30th year surveys were 50.5%, 44.4%, and 40.9%, respectively. The overall survival rates in the 10th, 20th, and 30th year surveys were 88.9%, 87.6%, and 85.7%, respectively. The surgical outcome of biliary atresia has markedly improved owing to the cooperation between Kasai portoenterostomy and liver transplantation. A comprehensive Japanese database of patients with biliary atresia (https://jbas.net/en/national-registration/) is now available.

INTRODUCTION

Biliary atresia (BA) is one of the most devastating biliary diseases affecting newborns and early infants. The uncorrectable type of BA that most patients fall into was completely fatal until 1959, when a corrective surgery, Kasai portoenterostomy (KP), was developed by Kasai. Kasai reported on KP in 1963 and 1968 in German and English, respectively.[1 2] By the early 1970s, KP became established as a standard procedure in Japan, leading to marked improvements in surgical outcomes for BA at several centers. However, the role of KP was not well understood in Western countries in those days. Some physicians mistakenly believed that all the reported successful cases of KP were due to misdiagnosis of neonatal hepatitis as BA and that KP was ineffective against BA. To promote understanding, Kasai proposed establishing the Japanese Biliary Atresia Society (JBAS) (The address of the Japanese site is "https://jbas.net", and the address of the English site is "https://jbas.net/en"). The JBAS was founded in 1975 to study all aspects of BA, and its primary role was to develop a clinical classification of BA. Currently, the JBAS has 103 member institutions, including most pediatric medical centers in Japan. As the diagnostic criteria and clinical classification of BA have been established, the usefulness of KP has gradually been recognized by pediatric surgeons worldwide, and it has become the surgical procedure of choice for BA since the late 1970s. Although KP became widespread, the surgical outcomes were far from satisfactory, even in Japan, in the 1980s. Therefore, in 1989, at the suggestion of Dr. Ryoji Ohi, the JBAS launched the Japanese Biliary Atresia Registry (JBAR) to investigate the epidemiology and etiology of BA and to improve surgical outcomes.[3]

Although the results of KP have improved over time, there have been many cases in which KP could not save lives, and in such cases, liver transplantation (LTx) was performed. In 1963, Starzl reported the first case of LTx, in which the recipient was a BA patient.[4] Although LTx results were initially poor, they improved considerably in the 1980s, and brain-dead donor transplants rapidly became widespread, especially in Europe and the USA.[5] Between 1988 and 1989, living donor partial LTx was performed in Brazil and Australia.[6 7] LTx has been clinically available in Japan since the early 1990s, and data on LTx recipients have been included in the JBAR since 1995.

In this review, the recent outcomes of the JBAR are presented.[8]

The JBAR system

The JBAR collects data using three types of questionnaires: an initial questionnaire, an LTx questionnaire, and a follow-up questionnaire. Pediatric surgeons from JBAS member institutions and hospitals affiliated with the Japanese Society of Pediatric Surgeons (JSPS) are responsible for registering patients and submitting data through an online system. It is assumed that every BA patient underwent surgery at one of these hospitals in Japan. From 1989 to 2023, 103 hospitals contributed to the JBAR, and pediatric surgeons who registered the initial questionnaire data were responsible for registering the follow-up data for the entire follow-up period of a patient.

The initial questionnaire was administered annually in June to collect data on new patients with BA who underwent KP or primary LTx between January and December of the previous year. The follow-up questionnaire was administered 2, 5, 10, 15, 20, 25, 30, 35, and 40 years after KP or primary LTx. The JBAR secretariat provides each facility with a list of patients to be followed up for that year, and the latest patient information at each time point is registered. If patients underwent LTx between January and December of the previous year, an LTx questionnaire was administered in June to collect recipient information. If LTx was performed in the same year as KP, the initial registration and LTx registration were conducted simultaneously in June of the following year.

As of 2023, 3951 patients had been registered, with 1688 undergoing LTx. The results of the registered cases are published annually in the Journal of the JSPS and on the JBAS website.

Initial questionnaire

From 1989 to 2023, the JBAR accumulated data from the initial questionnaires of 3951 patients with BA. The initial questionnaire consisted of 80 questions on family history, perinatal history, diagnosis, surgery, and outcomes.[3]

Patient background

Two-thirds of the patients were female, and over half had normal meconium and yellow stools. The mean birth weight was 2900 g (range: 824-4940 g) and the mean gestational age was 38.6 weeks (range: 24.0-42.0 weeks). Prenatal diagnosis (1991~) was possible in 5.5% (*n*=144/2605) of patients. Neonatal jaundice was not evident in approximately 30% (*n*=1030/3367) of the cases, and less than half needed phototherapy. The mean levels of total and direct bilirubin before the surgery were 9.3 mg/dL (range: 1.4-58.2 mg/dL) and 6.3 mg/dL (range: 0.4-40.5 mg/dL), respectively. The incidence of associated anomalies was lower than that of other congenital conditions. Relatively common anomalies associated with BA included polysplenia/asplenia (*n*=87, 2.2%), intestinal malrotation (*n*=80, 2.0%), and situs inversus (*n*=50, 1.3%).

The proportion of patients with BA associated with splenic malformation (SM) in the JBAR is significantly lower than that in Europe and the USA because of the high incidence of BA without SM in Japan. The frequency of BASM, calculated from reports that included both the frequency of BASM and the number of births in the population, ranged from 0.029 to 0.081 per 10 000 births in Europe and the USA, 0.013 per 10 000 births in Korea, and 0.061 per 10 000 births in New Zealand. Based on the number of registered cases in JBAR, the frequency in Japan was 0.023 per 10 000 births.[8–16] The actual incidence of BA associated with SM is not significantly different between Japan and Western countries.

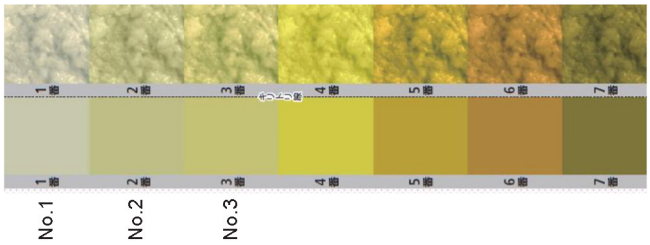

Figure 1 Stool color card. This card has been included in the Maternal and Child Health Handbook of Japan since 2012. If the stool color is similar to card numbers 1, 2, or 3, the infant's family is advised to visit a pediatrician or pediatric surgeon who uses this system. Figure from Gu *et al.* with permission of the Creative Commons CC-BY-NC-ND 4.0[17] .

Diagnostic modalities

Ultrasonography was the most common diagnostic tool, with 94% (*n*=3713/3951) of the patients undergoing ultrasonography. Hepato-biliary scintigraphy, serum bile acid, and duodenal fluid sampling were used to evaluate 50% or more of the patients. Preoperative CT or MRI was performed in less than 10% of cases.

Stool color card

In Japan, stool color cards were included in the Maternal and Child Health Handbook in 2012 to aid in the early diagnosis of BA. If the stool color is similar to card numbers 1, 2, or 3, the infant's family is advised to visit a pediatrician or pediatric surgeon using this system (figure 1).[17] The stool color cards comprise seven digital photographic images, and the stool color is inspected at 14 days and 1–4 months of age.[17] Of the 1177 cases registered between 2012 and 2023, color cards were obtained from 450 patients until 2023. Before 1 month, around 1 month, and after 1 month of age, 55% (*n*=121/220), 61% (*n*=156/255), and 87% (*n*=174/199) of the patients passed card numbers 1, 2 or 3 among cases with stool color cards number recorded, respectively. The rest of the patients passed stools of card numbers 4, 5, 6, or 7. The mean age at surgery was 70.6 days and 60.6 days in 2011 and 2023, respectively. The jaundice clearance rates were 57.8% (*n*=63/109) and 64.2% (*n*=52/81) in 2011 and 2023, respectively. Stool color cards could be effective for early diagnosis and potentially lead to improved surgical outcomes. However, currently, only approximately half of the registered patients have their stool color numbers recorded. Therefore, it is necessary to further promote the use of stool color cards.

Pathological bleeding

In this study, cases registered as abnormal bleeding in the JBAR were defined as bleeding thought to be caused by vitamin K deficiency. Vitamin K deficiency bleeding before KP was observed in 8.8% (*n*=335/3809) of patients, and intracranial hemorrhage (ICH) was the most frequent condition in 168 patients, followed by gastrointestinal hemorrhage in 101 patients, subcutaneous hemorrhage in 34 patients, and others in 58 patients. The average age

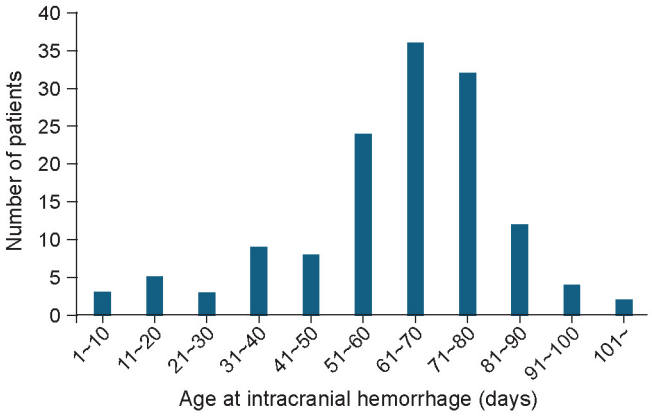

Figure 2 Age at intracranial hemorrhage among the registrants. The mean age at the onset of intracranial hemorrhage was 62.1 days, and approximately 80% cases occurred after day 51.

at ICH onset was 62.1 days old, and ICH onset occurred after day 51 in 110 of 138 cases (80%) (figure 2). Early diagnosis has a significant impact on the outcome of KP but is also crucial in preventing ICH.

Age at surgery

The mean age at surgery was 65.2 days, with a peak between 46 and 60 days. The age at surgery and jaundice clearance rate were correlated. Patients who underwent surgery before 80 days of age had better outcomes in jaundice clearance, which decreased significantly after 80 days (figure 3). In particular, patients who underwent surgery during the neonatal period showed the best jaundice clearance rate (almost 70%). The mean age at surgery has gradually decreased over time.

Surgical treatment and reconstruction procedures

KP was performed in 93% (*n*=3656/3951) of cases, hepaticoenterostomy was performed in 5% (*n*=211/3951) of patients, and primary LTx was performed in 0.5%

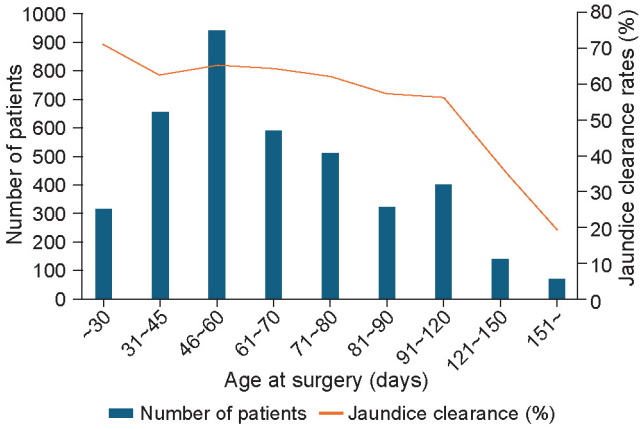

Figure 3 Age at Kasai portoenterostomy and jaundice clearance. There was a peak between 46 and 60 days old in the distribution of the age at surgery. Patients who underwent surgery during the neonatal period showed the best jaundice clearance.

(*n*=19/3951) of patients. Among the registrations regarding laparoscopic surgery that were started after 2018, the proportion of laparoscopic surgeries was 13.8% (*n*=73/529).

Registrants underwent various reconstruction procedures. Among them, the original Kasai, intestinal antireflex valve (IAV), and Suruga II procedures (total external biliary drainage) were common. The original Kasai procedure is also classified into two categories based on the length of the raised jejunum: the long Roux-en-Y procedure (LR), with a Roux-en-Y loop length more than 40 cm, and the short Roux-en-Y procedure (SR), with a loop length less than 40 cm. The IAV procedure includes intussusception and spur valves. There were 1843 cases of LR, 986 cases of SR, 682 cases of IAV, and 164 cases of Suruga II.

The jaundice clearance rates were 63% (*n*=1155/1843), 62% (*n*=607/986), 63% (*n*=429/682), and 55% (*n*=90/164) for LR, SR, IAV, and Suruga II, respectively. The incidence of cholangitis after LR, SR, IAV, and Suruga II was 42% (*n*=774/1843), 45% (*n*=447/986), 35% (*n*=242/682), and 46% (*n*=76/164), respectively. Among the four procedures, the incidence of cholangitis was the lowest in patients after IAV, although the jaundice clearance rate was similar, except for Suruga II.

KP revision, including hepatic hilar curettage, was performed in 452 patients, with jaundice clearance in 171 patients (38%).

Jaundice clearance and cholangitis according to the type of obstruction

The frequency of the type of obstruction varied greatly depending on the site of biliary obstruction: type I/I cyst, atresia of the common bile duct, 11.6% (*n*=452/3911); type II, atresia of the hepatic duct, 1.7% (*n*=66/3911); and type III, atresia at the hepatic hilum, 86.8% (*n*=3393/3911).[18] The jaundice clearance rates for type I, I cyst, II, and III were 70% (*n*=88/125), 79% (*n*=258/326), 72% (*n*=47/65), and 60% (*n*=2005/3356), respectively. The incidence of cholangitis in type I, I cyst, II, and III was 42% (*n*=53/125), 39% (*n*=127/326), 31% (*n*=20/65), and 42% (*n*=1418/3356), respectively. The incidence of cholangitis, including all types, was 50% (*n*=62/124) in 2000 and 44.4% (*n*=36/81) in 2023. The obstruction type was a significant prognostic factor following KP.[19]

Outcomes 1 year after the surgery

In the initial questionnaire, 57% (*n*=2218/3919) of patients were alive without jaundice, 13% (*n*=490/3919) had jaundice, 22% (*n*=858/3919) had undergone LTx, and 6% (*n*=218/3919) had died. The outcome trend in the initial questionnaire showed that the number of survivors following LTx markedly increased since the 1990s, whereas the number of deaths decreased significantly.

Follow-up questionnaire

The follow-up questionnaire focuses on the requirement of LTx, changes in liver function, development

Table 1 Follow-up data of native liver survivors

	Follow-up survery						
Follow-up data	2nd	5th	**10th**	**15th**	**20th**	**25th**	**30th**
Number of NLS	1855	1429	1040	729	446	234	79
Mean T-Bil (mg/dL)	1	1	1.1	1.2	1.1	1.5	1.6
Mean ALT (IU/L)	67.5	51.5	49.5	38.6	42.7	37.2	42.4
Mean γGTP (IU/L)	164.4	99.3	87.2	83	95.1	118.8	110.8
Normal Ch-E (%)	69	68	69	66	64	54	56
Esoph. Varices (%)	18	28	26	25	23	19	18
Hypersplenism (%)	18	29	35	31	28	30	25
Cholangitis (%)	21	20	14	19	23	28	23
Mentally delayed (%)	4	4	4	3	3	4	4
Physically delayed (%)	7	3	2	2	2	1	1

ALT, alanine aminotransferase; Ch-E, cholinesterase; Esoph. Varices, esophageal varices; NLS, native-liver-survival; T-Bil, total bilirubin; γGTP, γ-glutamyltranspeptidase.

of complications, developmental status (mental and physical), marriage, pregnancy, patient outcomes, *etc.*[3] Furthermore, from 2023 onwards, we added severity assessment items in line with "designated intractable diseases" under the national medical expense subsidy system in Japan to comprehensively grasp the severity of the disease. Follow-up is scheduled for the 2nd, 5th, 10th, 15th, 20th, 25th, 30th, 35th, and 40th years postsurgery.

Liver function tests in native liver survivors

The average liver function data for native liver survivors at the time of follow-up registration showed no tendency for alanine aminotransferase levels to worsen with age; however, total bilirubin levels increased slightly in the 25th and 30th years, and the proportion of normal cholinesterase values tended to decline from 60% to 50% (table 1).

Portal hypertension, cholangitis, and developmental status in native liver survivors

The frequency of both esophageal varices and hypersplenism tended to increase between the 1st year and the 5th or 10th year, but thereafter no major changes were observed, or there was a tendency for a slight decline. Cholangitis was somewhat less common in the 10th year (14%) but occurred in approximately 20% of cases at other times. Even in the 30th survey, 23% of native-liver survivors developed cholangitis. Despite this, their physical and mental development was generally within the normal range (table 1).

Native liver and overall survival rates

The native liver survival rates in the 10th, 20th, and 30th year surveys were 50.5%, 44.4%, and 40.9%, respectively. The overall survival rates in the 10th, 20th, and 30th year surveys were 88.9%, 87.6%, and 85.7%, respectively

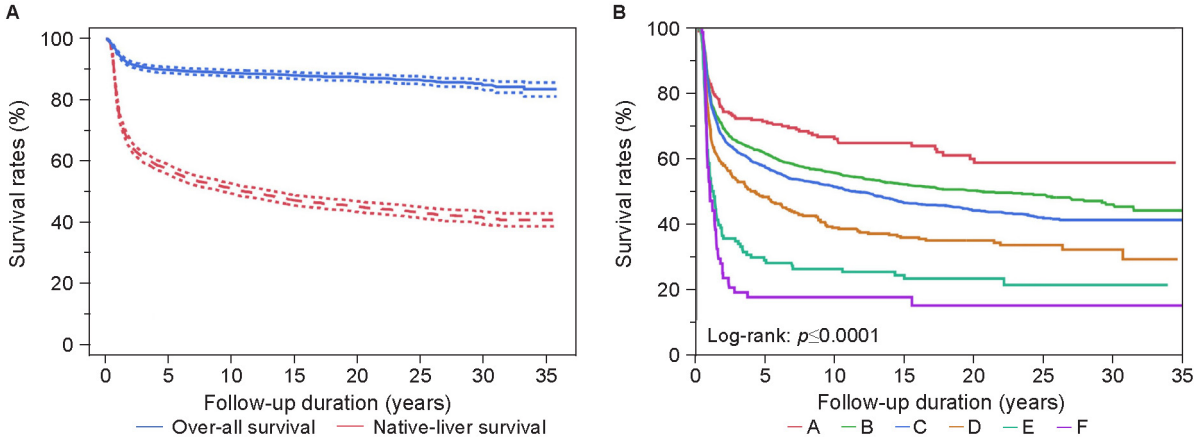

Figure 4 (A) Kaplan-Meier survival curves of the registrants. (B) Native liver survival curves according to age at surgery. Age at surgery (days): A, –30; B, 31–60; C, 61–90; D, 91–120; E, 121–150; F, 151–. The age at surgery had a significant impact on the long-term native liver survival rate.

(figure 4A). KP timing has been shown to significantly influence outcomes.[20] Patients who underwent surgery during the neonatal period had the most favorable prognosis (figure 4B).

Designated intractable diseases

At 30 years, the registry data showed that 65 and 79 patients survived with and without LTx, respectively. Of the 79 native liver survivors, 16 had jaundice and 59 did not. 28 of the native liver patients were married, and 23 became pregnant. Regarding the severity assessment of designated intractable diseases in the 20th year survey of native liver survivors, 56% (n=18/32) were assigned to level 0 (no signs or symptoms of BA), 28% (n=9/32) to level 1 (mild signs or symptoms of BA, no need for LTx), 6% (n=2/32) to level 2 (moderate signs or symptoms of BA but no need for prompt LTx), 3% (n=1/32) to level 3 (severe signs or symptoms of BA, requiring LTx), and the remaining 6% (n=2/32) had no assessment recorded. As mentioned previously, by the age of 20 years, 55.6% of patients required LTx or died; therefore, it is likely that approximately 60% of patients would have reached level 2 or 3 at the time of the 20th year survey. The condition of adult patients with BA is gradually becoming clearer. However, as the number of long-term survivors remains small, continuous monitoring is needed.

LTx questionnaire

A total of 26 questions were asked in the LTx questionnaire, including questions on recipient and donor details, types of transplantation, liver function tests, complications, immunosuppression protocols, and patient outcomes.[3] A total of 1688 patients have undergone LTx in the JBAR until 2023. LTx became more common in Japan in the late 1990s, with approximately 40% of registered BA patients requiring transplantation over the last 30 years (figure 5).

Donor source

Only 39 patients received grafts from deceased donors. Most LTx procedures (especially in pediatric cases) rely heavily on living donors, typically the parents of the recipients, highlighting the need for a stronger deceased donation system in Japan.

Patient survival at LTx registration

Of the 1688 transplant registrants, 1564 (93%) survived at registration. However, living donors are the primary source; the establishment of deceased donor transplantations is awaited in Japan.

Current status and future perspectives of the JBAR

Since 1989, epidemiological and clinical information on BA in Japan has been accumulated in the JBAR. This information is already being distributed worldwide via the JBAS website, making it available to everyone interested in BA, including medical professionals, researchers, and patients and their families.[8]

The clinical practice guidelines (CPGs) for BA were created in 2018 by the JBAS, and an English abbreviated

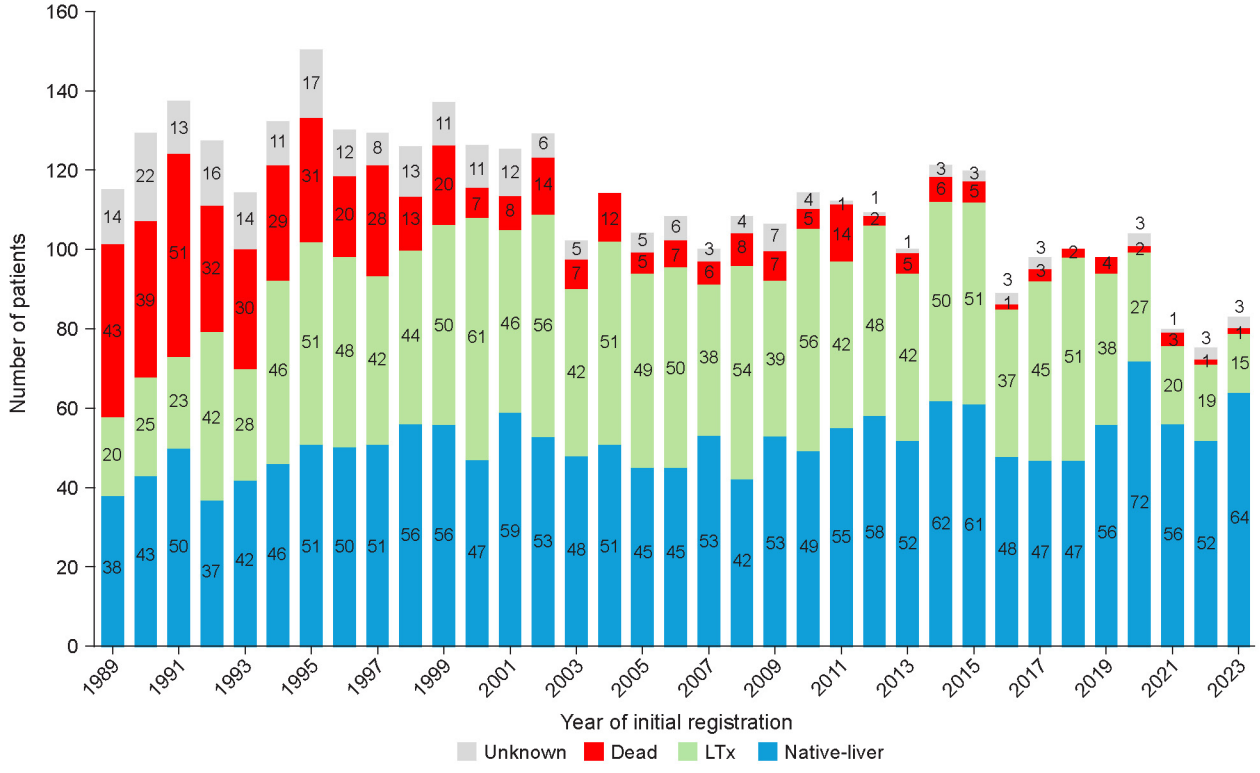

Figure 5 The current status of patients who entered the JBAR in each year is shown. JABR, Japanese Biliary Atresia Registry; LTx, liver transplantation.

version of the CPGs was published in 2020.[21] The JBAR data greatly strengthened the evidence base for the CPGs. The CPGs were recently revised, and the latest JBAR data were abundantly featured.

While the JBAR is accumulating information on long-term survivors every year, the number of patients lost to follow-up is also increasing annually, making it difficult to obtain a complete picture of patients, especially older adult patients. This tendency is partly due to the current registration rules, in which follow-up registration is stipulated as the responsibility of the institution where the initial registration took place, most of which are pediatric medical centers. These rules create obstacles to follow-up registration as patients grow older, undergo transplantation, or move elsewhere. Amending the regulations to allow the follow-up facility to take over registration, if the site is changed, may be beneficial.

The Japan Liver Transplant Society runs a registration system in parallel for the follow-up of liver transplant patients. Therefore, by deepening the collaboration between the JBAS and Japan Liver Transplant Society, it is expected that the burden on registration staff at each facility will be reduced and more efficient, and detailed research will be possible, targeting a larger number of liver transplant patients.

In addition, the JBAR plans to observe patients for up to 40 years after KP, and enrollment for the 35th year is currently underway. Because the number of BA patients aged 50 years or older is gradually increasing in Japan, it may be necessary to extend the follow-up period of the JBAR to 50 years, 60 years, or even longer.

In addition, clinical studies using the JBAR data have been available to the public from 2021, and various analyses using the JBAR data have been planned. Detailed analyses are expected to advance research from various perspectives, such as elucidating pathology and predicting prognosis.

It is our sincere hope that further continuation of the JBAR will help to overcome the various challenges faced by BA patients and their families.

CONCLUSION

Between 1989 and 2023, 3951 patients had been registered in the JBAR. Among them, 1688 underwent LTx. Follow-up questionnaires for up to the 40th year are ongoing. Currently, a large Japanese database (https://jbas.net/en/national-registration/) is available.

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Competing interests MN and the Japanese biliary atresia society were supported by The Health and Labor Sciences Research Grants (22FC1014).

Patient consent for publication Not applicable.

Ethics approval The study protocol for the JBAR has been approved by the Clinical Research Ethics Board of Tohoku University Graduate School of Medicine (no. 2024-1-242).

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Data availability statement Data are available on reasonable request.

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