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How to evaluate diagnosis and management of biliary atresia in the era of liver Transplantation in China

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ABSTRACT

Background Biliary atresia (BA) is one of the most challenging hepatobiliary diseases in children. Notwithstanding the reasonable outcome of liver transplantation (LT), portoenteral anastomosis (Kasai operation) is still the first choice of treatment for patients with BA. How to improve the survival rate of the patients with BA in the era of LT is a new challenge.

Data sources Based on recent original publications and the experience with the BA in China, we review many factors that influence BA survival situation, including early diagnosis and screen plan, defects of early treatment, Kasai operation, and LT and indicate present questions about BA diagnosis and treatment in China.

Results BA diagnosis may also be delayed due to insufficient understanding of BA and lack of jaundice monitoring methods at different levels of the hospitals in China. Further education of the physicians at smaller city hospitals about BA, neonatal jaundice and cholestasis would be helpful in improving early diagnosis of BA. Early surgical intervention is still the only guarantee to improve the survival rate of BA with native liver.

Conclusions In the era of LT, especially in China, Kasai operation can provide waiting time for living donor LT and improve the success rate of LT and minimize the rapid deterioration of liver function of the children with BA and decrease the mortality in patients with BA.

INTRODUCTION

Biliary atresia (BA) is one of the most challenging hepatobiliary diseases in children. BA is not a common disorder. The estimates of national prevalence in UK and France ranged from 1 in 17 000 to 1 in 19 000 live births.[1] In China, it is estimated to be 1 in 7000 in the Tianjin region.[2] Following the opening of the second child policy, new birth population estimated 20–30 million per year, so we figure down almost 3000–4000 new BA cases in China every year. However, the outcome of current treatment is not satisfactory. It was poor for 2 years native liver survival after Kasai procedure (less than 50%) and the rate of performing Kasai procedure in China. Liver transplantation (LT) for patients with BA is a revolutionary progress, providing a new possibility for patients with BA with end stage liver failure. It is quite clear that if patients with BA do not have the Kasai procedure, they will die without having transplantation. Since the first successful LT for BA in 1963, LT techniques have become advanced in Europe, the USA, and Japan. It also had been established in Mainland China. Last year, there were 700 patients with BA who received LT in China. Notwithstanding the reasonable outcome of LT, portoenteral anastomosis (Kasai operation) is still the first choice of treatment for children with BA.

How to improve the survival rate of the patients with BA in the era of LT is a new challenge. In China, nowadays, excessive publicity of the advantages of LT by media and some of the surgeons and incorrect information on the internet lead to parents' thinking of the poor effectiveness of Kasai procedure. So the parents of patients with BA prefer to have their children accept LT first. The percentage of LT without prior Kasai procedure was higher than other countries, especially Japan (table 1). However, in fact, the Kasai procedure prolongs the native liver survival and improves the survival rate of patients with BA.[3] It is clearly recommended that in patients with BA, the Kasai procedure should be performed first unless there is an advanced liver failure at presentation. Then, the LT should be performed for those who have either a failed Kasai operation or progressed end stage liver disease. It is very important for the patients with BA who had suitable time to perform Kasai procedure. Based on the basic and clinical research of BA, there are some considerations that need to be addressed early about BA to complete Kasai procedure: (1) early screening and diagnosis of BA enables early Kasai procedure and good results. A French BA research center showed that[4] the jaundice clearance rate was 51.5%, 43.1%, and 31.6% when the Kasai procedure was done at 1 month, 2 months, or 3 months after birth, respectively. Furthermore, a study from Switzerland confirmed that the

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Table 1 The rate of liver transplantation without prior Kasai procedure in the Chinese cities and other countries

Author	Year	Area	Time (year)	Patients (cases)	Liver transplantation without prior Kasai procedure (%)
China					
M M Tiao	2008	Taiwan	1996–2004	106	46 (43.4%)
Chunbao Guo	2010	Sichuan	2006–2009	22	13 (59.1%)
Qiao Wang	2013	Sichuan	2008–2011	28	18 (64.3%)
Shanni Li	2016	Tianjin	2006–2014	150	60 (40%)
Other countries					
S P Alexopoulos	2012	USA	1995–2008	134	22 (16.4%)
J S Neto	2015	Brazil	1995–2013	347	141 (40.6%)
T Kitajima	2017	Japan	2005–2016	173	5 (2.9%)

jaundice clearance was only 10% if the Kasai operation was performed beyond 75 days,[5] and the authors proposed that the most important measure to improve the prognosis in patients with BA was to decrease the age of Kasai operation. (2) Failure to recognize BA at an early age delays the diagnosis of BA. Song *et al*[6] from Shanghai, China reported 498 patients whose first presentation was 30 days but Kasai procedure was performed after 70 days in China. In fact, the patients sought help early, but different local physicians lacked the awareness of BA. As we know, Kasai procedure only can release extrahepatic biliary obstruction, yet the obstruction of intrahepatic biliary still existed. In addition, it is essential to control intrahepatic biliary tract inflammation and infection with intravenous antibiotics and corticosteroids after Kasai procedure.[7] So far, there were standardized postoperative management protocols which had been published in Chinese Journal of Pediatrics.[8 9] In these protocols, including antibiotics, corticosteroids, and hepatoprotective cholagogue should be used for a longer time after Kasai procedure. For corticosteroids, the recommended plan 1 was taking prednisolone after recovery of intestinal function, 4 mg/kg/day for 4 weeks, per os, once a day, taken in the morning, which reduced to 2 mg/kg/day for 4 weeks, followed by reduction to 1 mg/kg/day and then stop it after 4 weeks. Plan 2 was taking methylprednisolone after recovery of intestinal function post Kasai procedure, 10, 8, 6, 5, 4, 3, 2 mg/kg/day for total 7 days, ivgtt, which changed to 2 mg/kg/day for 4 weeks, per os, followed by reduction to 1 mg/kg/day (per os) and then stop it after 4 weeks. For antibiotics, plan 1 was taking cefoperazone 60–80 mg/kg/day ivgtt for 7 days, two times a day, which changed to meropenem 25 mg/kg/day, 8 hours, for 4 weeks and then changed as oral antibiotic for 3 months. Plan 2 was taking ceftriaxone 20–80 mg/kg/day ivgtt for 7 days, once a day, followed by change to meropenem 25 mg/kg for 4 weeks, 8 hours and then changed as oral antibiotic for 3 months. In contrast, the recommendation for standard use of corticosteroids post Kasai procedure was not valid based on the most recent placebo-controlled US trial. Both pediatric surgeon and liver transplant surgeons agree that the first choice for the patients with BA is Kasai procedure, and LT is reserved for children with BA with end stage liver failure after Kasai procedure. Our data showed that there are significant differences between Kasai and non-Kasai group in the appearance of jaundice, Kasai procedure age, transplantation age, and operative time (table 2). Kasai procedure can delay the waiting time of patients with BA for LT. (3) In most people's mind in China, patients with BA might have poorer prognosis, especially the patients with BA associated with heart malformations were more likely to suffer from hepatopulmonary syndrome and had higher rates of mortality after Kasai operation. For patients with non-Kasai procedure, there were different reasons including physicians and the thought about economical situation. Although they thought about the family's economical situation and the prognosis, sometimes, parents of patients with BA would choose the cholangiogram to confirm the BA diagnosis rather than to perform the Kasai operation. Physicians, who came from local hospital (not Children's hospital), also recognized that patients with BA would not survive longer after Kasai procedure, so they advised them to give up the Kasai procedure and the whole treatment or to take the earlier LT option.[10] There were some reasons that influenced the results of BA treatment in China, and we list these situations below.

The work for early diagnosis and screening program in China

BA accorded with the basic requirement factors of early screening, that is, patients with BA have serious health problems without clear clinical symptoms and early diagnosis can improve the therapeutic outcome and prognosis.[11] Many medical practitioners attempted to diagnosis BA by simple, convenient, inexpensive, and highly sensitive methods.[12] For example, Stool color card, B-ultrasonography, and serum conjugated bilirubin level measurement are effective screening methods which have been widely used in the USA, Canada, Japan, Britain, France, and other countries.[13] While stool color card's efficacy has been proven for early diagnosis of BA in Canada and Japan, the routine BA screening was not included in US neonatal disease screening protocol. In

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Table 2 Two groups were compared with present time of jaundice, transplantation age and operative time

Group	Cases	the appearanceof jaundice (days)	Kasai age (days)	Transplantation age (days)	Operative time of Tx (hours)
Non-Kasai	31	31 (0–60)	–	310.90±210.15	9.29±1.41
Kasai	55	33 (0–75)	66±25.48	549.49±494.46	8.65±1.43
P value	–	0.974	–	0.008*	0.047*

Non-Kasai group: The average age of liver transplantation was 310.9 days (174–1285) and average transplantation time 9 hours 17 min (6 hours to 12 hours 15 min).
Kasai group: The present time of Jaundice is 28 days, but the Kasai time is 70.39 days. The average age of liver transplantation was 549.5 days (173–3100), and average transplantation time 8 hours 40 min (5 hours 15 min–14 hours). Significant differences existed in transplantation age and surgical duration (p<0.05), and present time of jaundice was not statistically significant.
*P<0.05, significant differences.

Britain, serum conjugated bilirubin is widely screened in cases of neonatal jaundice. The stool color card was already included in neonatal disease screening system in South America and some Asian countries and regions. Serum conjugated bilirubin was effective to instigate earlier referral, and in most of countries cited like the USA and Britain or France, and there were no standardized policy for conjugated bilirubin testing. Yet, BA screening plan has not been included in the newborn screening program in most provinces of our country. In China, Shenzhen was the first region to put the Stool color card into neonatal healthy checking handbook which printed color stool card and then used it to screen neonatal jaundice disorder. The cost-effectiveness of the stool color card screening program in Shenzhen region was not so expensive, which was about 10 yuan ($1.6) for each healthy checking handbook. The families themselves did the primary screening at home, and when they found the stool color change abnormal, they would contact with physicians from China Shenzhen Children's Hospital. They printed the telephone number and QQ number on this card. The physicians need to do different diagnoses of BA using conjugated bilirubin testing and B-US. So, a good suggestion to the different city health bureaus and a comprehensive plan by the medical administration at all levels hospital are urgently needed in the management of neonatal jaundice and for the early diagnosis and treatment of BA. And further education of about BA-related basic information should be provided to physicians who worked in the small-town hospitals.

Neonatal jaundice and cholestasis would be helpful in improving the early diagnosis of BA. Jaundice, clay stool, and dark urine colors are the main clinical symptoms of BA. Our team have travelled almost 10 cities (including Hefei, Chengdu, Xi'an, Nanchang, Changsha, Shijiazhuang, Yinchuan, Urumchi, Changchun, Guiyang, and so on) to popularize the knowledge about BA. However, most of the patients did not present all symptoms, and the diagnosis may be delayed when the symptoms appeared in full. Therefore, to establish a screening process for BA is very important. The national BA seminar was held in Tianjin region from 2012 till 2017, and experts from various regions of China discussed and approved the basic steps of early screening for BA, which are as follows: (1) advocated a health check-up at 1 month after birth and to perform the hepatobiliary B-ultrasound if indicated; (2) if there was a persistent jaundice, the liver function tests including conjugated bilirubin, γ-GT, and ALT should be tested; (3) a handbook of physical examination was recommended, stool color cards should be provided, and the help of pediatric surgeons should be sought when the stool color was abnormal.[14] The above screening recommendations should be part of the newborn health handbooks, and the parents and the physicians can detect the disorder easily, and the patients with BA should opt Kasai procedure earlier. So far, the healthcare providers in Tianjin, Shenzhen, Shanghai, Wuhan, LiaoNing, ChenDu, and HangZhou have widely used stool color card to make the screen work.

The reason why BA diagnosis was delayed may also be due to insufficient understanding of BA and lack of jaundice monitoring methods at different levels of the hospitals. At the time of transferring to the pediatric medical center, patients with BA often have developed severe cirrhosis at the time of presentation, further prohibiting Kasai operation completed during operation. There are also incidences where physicians were discouraged by the poor outcome of Kasai operation and suggested to the patient's family to give up Kasai procedure, which further narrowed the window of opportunity for Kasai operation outcome. Another reason of low rate Kasai procedure was the relatively high cost of diagnosis and treatment of BA and the cost of the repeated cholangitis. Some families cannot afford this expenditure and want to give up the treatment. Usually, the medical costs for BA diagnosis is almost 10 000–15 000 yuan ($1600–2400) including blood liver function, B-US, open liver biopsy, and operative cholangiograms. If Kasai operation is needed, the family should pay extra cost of 25 000 yuan ($4032). In recent years, the China public health insurance program would cover about 40% medical costs for BA diagnosis and treatment, and in the coastal area, the reimbursement rate reach to 60%–80%. On 28 March 2013, Health News reported that China's first funded project of Kasai procedure for children with BA in poor areas had started in Shanghai region. The cost of the Kasai procedure was provided by the central government (only for 100 patients with BA). On 1 July 2013, Chinese Assistance

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Foundation of Organ Transplantation started the rescue plan of living-related LT for the children with BA from poor families in Tianjin. They have funded close to 100 living liver transplant surgeries to patients with BA in our country. The implementation of these aid projects for patients with BA undoubtedly benefits the vulnerable groups in China.

Defects of early diagnosis and treatment

The obstacle of diagnosing BA lies in the fact that: (1) the incidence of BA is relatively low. Only 2%–15% of neonatal jaundice persist for more than 2 weeks after birth; for every 500 children with jaundice, 1–2 final diagnosis of BA was made; the awareness of BA among pediatricians and child healthcare physicians was not high. (2) Lack of simple, convenient, and effective confirming diagnosis tools; Stool color cards as a screening tool is very useful. Practical means of investigations at present include a fecal card and serum direct bilirubin blood test. The specificity of stool color cards as a screening tool for patients with BA is very high and fewer were false negative. The direct bilirubin test for BA has high negative predictive value. (3) Neonatals who present to the specialist physicians were often too late, usually 8 weeks after delivery rather than 3–4 weeks postnatal, and they missed the optimal time to identify BA.[15]

American Academy of Pediatrics (AAP) recommended to test the serum total and direct bilirubin in all infants with persistence jaundice beyond 3 weeks of age.[16] In practice, it is difficult to implement these measures due to various reasons. Currently, this recommendation is often questioned by pediatricians, mainly due to the relative low incidence of BA.[17] About 95% of BA with appearance of jaundice in 3 weeks after birth were considered "healthy" by parents and physicians; if the patients with jaundice visited the hospital in 3–4 weeks after birth-persistent jaundice, some children with neonatal cholestasis may be diagnosed with BA early. Most parents follow the doctor's advice for subsequent visits, but many doctors do not pay attention to the follow-up time of jaundice. The majority of them were over 6 weeks after birth; for mild jaundice, doctors always choose nonoperative treatment, without timely communication with pediatric hepatobiliary surgeons. Therefore, North American Society of Pediatric Gastrointestinal and Nutrition underlined the need for basic knowledge education of the medical staff training in the field of BA, if the patients with persistent jaundice should undergo specialized investigations by pediatric hepatobiliary surgeons and not by the internists,[18] and there were same issues in China. The baby with jaundice in his first month was mostly considered to be due to breast milk feeding. They still waited until the jaundice got worse and they would ask hepatobiliary surgeons to do an operative cholangiogram and differentiation from other causes of intrahepatic cholestasis. Song *et al*[6] reported 498 patients from Shanghai region in China whose first consultation is 30 days but surgery was not performed until 70 days. Zhan *et al*[19] collected the data of 851 BA jaundice patients first present in the hospital was 28–32 days, the Kasai procedure is still delay to 70.39 days.

Kasai procedure and liver transplantation

The postoperative care of patients with BA was done by pediatric surgeons, especially pediatric liver specialists in China. So, there were more problems present after Kasai operation. The majority of BA after Kasai operation will be accompanied by a relatively lengthy process of recovery, including antibiotics, corticosteroids, and cholagogue medications.[20] Some patients even used traditional Chinese medicine treatment. Most patients with BA have only one chance for Kasai procedure and do not have chances of a reoperation in future. If these measures were proven ineffective, the patients may need to give up treatment after surgery, as other patients with BA are waiting for the LT. Ascending cholangitis or recurrent later cholangitis after Kasai portoenterostomy blocked the biliary juice drainage. Reoperation or wanting for the LT has been the controversial titles among pediatric surgeons and liver transplant surgeons. Reoperation for BA was a major undertaking, and its long-term prognosis was uncertain. The decision was difficult to make for both doctors and parents. If the surgeon chooses reoperation, he/she must fully understand the physiological state of the patient, including the liver function and possible liver fibrosis level. For those who have mild liver fibrosis, patients can choose reoperation; for those with severe liver cirrhosis, they must be careful to select re-do Kasai portoenterostomy. It may be better to prepare the patients with BA for early LT.[21] In the era of LT, BA reoperation can provide waiting time for donor liver and improve the success rate of LT and minimize the rapid deterioration of liver function of the children with BA and further decrease the mortality. Therefore, an evaluation of liver before reoperation is crucial.[22] The opponents of reoperation of BA argue that nearly 70% of the children with BA in the future need LT and it is meaningless to complete reoperation. Another rather radical view of the management is that the children with BA should wait for a LT directly without going through Kasai operation. However, the statistics data show that the children with BA who have LT after Kasai procedure achieve greater rate of survival compared to the non-Kasai patients. The clinical data showed that Kasai operation should prolong the native liver survival time and children with BA can grow up and wait for LT.[23]

In the USA, since the advent of LT in 1960s, there had been about 13 000 LTs. The cases increased by a few hundred each year, and the long-term survival was obviously better than that of adult patients. In Japan, from November 1989 to December 2015, 2085 patients with BA underwent LDLT, and the rates of 1-year, 5-year, 10-year, 15-year, and 20-year graft survival for the patients with BA were high.[24] In addition to the technical progress, there were also associated change of the primary disease of pediatric recipients of LT, that is, the main

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indications for LT in children are congenital and metabolic disease.[25] Besides, the 2016 annual report from the Scientific Registry of Transplant Recipients demonstrated 90.8% 1-year and 82.7% 5-year graft survival among pediatric liver transplant recipients.[26] LT in our country started late; there were about 540 pediatric living donor LT operations in mainland China in 2011, accounting for 2.59% of the total case load of LT. Sixty-eight per cent of pediatric liver transplants in China were for BA. The overall 5-year transplant survival for patients with BA is 93.3%. Living donor LT in children accounted for 66.3% of pediatric LT and 21.1% of the total number of living-related LT (China Liver Transplant Registry: http://www.cltr.org). Although there are many children's medical centers performing Kasai procedure for children with BA, the outcome was varied. There was no detailed statistics documenting the 3-year and 5-year survival rates of the native liver for patients with BA; so we need a large, multicenter, retrospective analysis to assess the results of BA treatment in our country and establish a good follow-up mechanism. LT as a treatment of BA is revolutionary, especially when living-related donor LT technology provides new source of donors. But in China, nowadays, the cost of LT was not comparable with the current economic level of development. Even in the developed countries such as Europe, America, and Japan, doctors still advocate the children with BA to undergo Kasai operation first, improving the survival rate of the native liver, until such time when terminal liver failure eventuates, and then consider a LT. For most of the children with BA, they only undergo Kasai procedure once; therefore, whether exploratory surgery or laparotomy operation, the pediatric surgeon should carry it out and should not it give up, as this is the only survival opportunity for patients with BA. The technology of LT is much more advanced now,[24] and the pediatric surgeons should have more positive attitude towards the treatment of BA.

In China, although the rate of Kasai procedure was about 90% in most of the cities (table 3), the rate of LT with prior Kasai procedure is only 38.1%.[27] There were some possible reasons as follows: (1) There were not too many centers which can perform Kasai procedure, and most patients with BA had no chance to accept the operation. (2) The rate of native liver survival post-Kasai was about 50%, so some of the patients with BA do not need to accept LT. (3) Some patients with BA died or were lost during follow-up before they accepted LT.

CONCLUSION

In conclusion, in the era of LT, we must better understand the diagnosis and treatment of BA in some instances. Early Kasai procedure is still the only guarantee to improve the survival rate of BA with native liver. For older children or those with failed Kasai surgery, LT can save the life. So far, Shanghai has established a national registry system for BA to accrue and assess outcome data. This would be a good beginning for the diagnosis, treatment, progresses, and survival situation of patients with BA.

Table 3 The data of Kasai procedures performed by cities of China and other countries

Author, Year	Area	Time (year)	Patients (Cases)	Kasai rate	The age of surgery (days)	Jaundice clearance rate (%)	Cholangitis incidence (%)	2/4 year native liver survival rate (%)	5/10 year native liver survival rate (%)
China									
Chunqiang, Dong, 2012	Guangxi	2007–2011	126	100	85	62.5	30.4	61.8 –	–
Haiwei Lin, 2012	Beijing	2005–2010	312	64	75	50	57	56 49	–
Zai Song, 2014	Shanghai	2004–2010	476	100	–	–	–	51.5 –	–
Lee M, 2016	Taiwan	1997–2010	513	89	46.4	–	–	81.1	–
Xue Sun, 2017	Shanxi	2009–2015	129	100	–	67	73.6	36.3 –	–
Other countries									
Nio, 2003	Japan	1989–1999	1381	83.5	–	57	–	–	60 –
Shneider, 2006	USA	1997–2000	104	100	61	38	–	56 –	–
Wildhaber, 2008	Switzerland	1994–2004	48	89.6	68	37	–	– 37.4	–
Schreiber, 2010	Canada	1992–2002	230	90.0	64	39	–	–	–
Leonhardt, 2011	Germany	2001–2005	183	86.9	57	20	–	–	18 –
Davenport, 2011	UK	1999–2009	443	95.7	54	38	–	–	56 49
Chardot, 2013	France	1986–2009	1107	94.3	59	36	–	–	46 40

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CAS academician Jinzhe Zhang[28] says: "To recognize the relationship between treatment of BA and LT means that the medical level of pediatric specialist surgeon as well as pediatrics in many ways is not only improved in the level of pediatric recognized, but a medical time progress in pediatrics traditional mind." This statement is a good summary of the status of BA, the existing problems and the future development direction of diagnosis, and treatment of BA. The diagnosis and treatment of BA is the duty of all pediatric medical workers, especially in the era of more advanced LT technology. Pediatric specialist surgeons of our generation should work hard, have positive enterprising attitude, and give the children with BA new hope of survival. We firmly believe that through our continuous efforts and in-depth study, miracles will happen in the area of the treatment of BA in a few years.

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Competing interests None declared.

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