



# Nutritional intervention and efficacy analysis of children with biliary atresia after Kasai portoenterostomy

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**Background:** Biliary atresia (BA) is the most common cause of cholestatic cirrhosis in children, and liver transplantation (LT) is the sole treatment for this disease. However, a young age and light weight increase the difficulty and risk of LT. Currently, Kasai portoenterostomy (KPE) is the preferred treatment for children with BA to buy time while waiting for a donor liver to become available. The pretransplant nutritional status is the key factor determining post-surgery quality of survival. Therefore, in this study, we followed 40 children with BA who survived with autologous livers after KPE from 2010 to 2017. Individualized nutritional guidance was given, and the post-KPE nutritional status of the children and the jaundice clearance rate were evaluated to better formulate follow-up and nutritional intervention strategies.

**Methods:** The multidisciplinary team (MDT) carried out regular physical examinations of the children after KPE and supplemented them with a physiological dose of vitamin D (400–800 IU). In addition to feeding guidance, children with malnutrition were given medium-chain triglyceride (MCT)-enriched nutritional formula milk. The children's body weights, body lengths, head circumferences, liver functions, bilirubin levels and plasma vitamin A, D and E levels were measured. The Z-score of physique and the percentiles were calculated using the anthropometric calculator of the WHO Anthro software, and descriptive statistics were evaluated. An independent-samples t-test was used to compare pre- and post-KPE indicators and vitamins.

**Results:** A total of 40 children were included in the study. Their jaundice clearance rate was 56% within 3 months after KPE. Among them, 25 children had good compliance and were followed up at least twice. The patient ages at the last follow-up were 13.38±21.70 months. The male-to-female ratio was 24:16. The mean Z-score and percentiles of body weight and body length/height at the last follow-up for the 40 children were weight for age (WFA) (-1.49±1.54, 26.53±25.91) and length for age (LFA) (-1.39±1.16, 18.93±18.96, respectively). A total of 17.5% (7/40) of the children were emaciated, and 22.5% (9/40) had a short stature. The vitamin A, D and E levels were 0.99±0.29 µmol/L (marginal level), 34.12±16.71 µmol/L and 12.82±1.53 µg/mL, respectively, which did not differ compared to those of children of the same ages from the outpatient children's health clinic. The prealbumin level was 150.82±47.48 mg/L (lower than the normal level), and the albumin level was 40.67±4.76 g/L (normal range).

**Conclusions:** The jaundice clearance rate was 56% (14/25) for this group of children with BA within 3 months after KPE. At the last follow-up (13.38±21.70 months), the emaciation rate was approximately 17.5% (7/40), and the short stature rate was 22.5% (9/40), which reflected more prominent long-term malnutrition among the BA children. Multidisciplinary cooperation is very important for long-term nutrition management of BA children after KPE.

**Keywords:** Biliary atresia (BA); Kasai; nutritional intervention; fat-soluble vitamin; malnutrition

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## Introduction

Biliary atresia (BA) is a disease involving partial or total obstruction of the intrahepatic and extrahepatic bile ducts, which leads to cholestatic cirrhosis and ultimately liver failure (1). The etiology remains unclear but may be related to genes, viral infection, toxins, chronic inflammation or immune-mediated bile duct injury (2). Without surgery, most children die within a year, and liver transplantation (LT) is the only treatment for the disease. Early diagnosis and timely Kasai portoenterostomy (KPE) are the preferred treatment for BA (3). However, even when KPE is performed immediately after birth, children with end-stage liver disease will eventually require LT (4). The liver is the major organ involved in nutrient metabolism. Although bile flow is established after surgery in BA children, malnutrition still occurs easily. Children with a high risk of malnutrition are more prone to cholangitis after KPE, and the jaundice clearance rate is lower than that of children with a low nutritional risk; thus, these factors affect the 2-year survival rate of children with autologous livers after KPE (5). A recent study showed that severe or persistent malnutrition after KPE was one indication of a need for LT. Ideal pre-surgery management should actively optimize nutrition, growth and development of these children (6).

To study the nutritional status and strengthen the nutritional management of children with BA after KPE, this study conducted regular follow-ups of children with BA after KPE in the Department of Neonatal Surgery of our hospital over the last 7 years. We studied their post-KPE jaundice clearance status, physical development status and plasma levels of fat-soluble vitamins to better develop strategies for follow-up visits and nutritional intervention and to improve the long-term survival and quality of life of children with BA.

## Methods

### *Research subjects*

The study subjects are children with BA admitted to the Department of Neonatal Surgery of our hospital

who survived with their autologous livers after KPE. Their physical status and fat-soluble vitamin levels in the peripheral blood were dynamically monitored to evaluate the post-KPE liver function, bilirubin level and nutritional status.

### *Research methods*

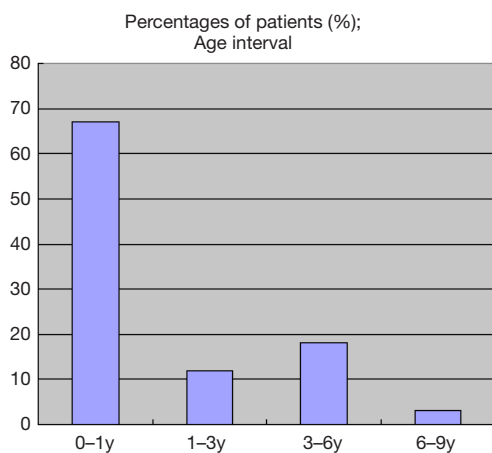
Patients with BA after KPE were followed up in the multidisciplinary postoperative follow-up clinic (MDT) and evaluated by neonatal surgeons, gastroenterologists and clinical nutritionists. The follow-up time usually was 1, 2, 3, 4, 5, 6, 9 and 12 months after KPE, and a re-examination was conducted annually after the age of 1 year. This study was reviewed and approved by the Guangzhou Women and Children Medical Research Ethics Committee, and the approval number was 26001.

**Nutritional intervention:** a simple dietary survey was conducted to assess the nutritional intake, and individualized nutritional guidance was given to the children according to their nutritional statuses. Because children with BA are at high risk of vitamin D deficiency, all children were required to take a physiological dose of a vitamin D supplement (400–800 IU) (7,8). In addition to feeding guidance, children with malnutrition were given a medium-chain triglyceride (MCT)-enriched nutritional formulation (9).

**Physical examination:** the height/length, weight and head circumference were measured by referring to the physical measurement method of Zhu Futang's Practical Pediatrics (the 8th edition). The weight of the infants and young children aged 1 to 3 years was measured with the electronic infant scale (seca376), whereas the weight of children aged 3 to 17 years was measured using the electronic standing scale (seca704s), which provided an accurate reading to 0.01 kg. The length of children under 3 years old was measured in the recumbent position (recumbent length) using the infant measuring bed (seca416). The height of children over 3 years old was measured using an electronic standing scale (seca704s), and the reading was recorded to within 0.1 cm. For children under 3 years old, the head circumference was measured using a seca201 measuring

ruler, and the reading was recorded to within 0.1 cm. The above Seca series instruments were purchased from Seca Company, Germany. At each clinic visit, the growth curves were plotted to assess growth and development trends.

Lab tests: regular checks of liver function, prealbumin and plasma levels of fat-soluble vitamins A, D and E were performed. Liver function was measured using a turbidimetric immunoassay. The fat-soluble vitamins were detected by chemiluminescence. Jaundice clearance 3 months after KPE is defined as a total bilirubin (TBIL) level <20 µmol.



**Figure 1** The age distribution of children followed up after KPE.

Statistical method: The Z-score of physique and the percentiles were calculated using the anthropometric calculator of the WHO Anthro software, and descriptive statistics were evaluated. An independent-samples *t*-test was used to compare pre- and post-KPE indicators and vitamins, with  $\alpha < 0.05$  set as the statistical significance level.

**Results**

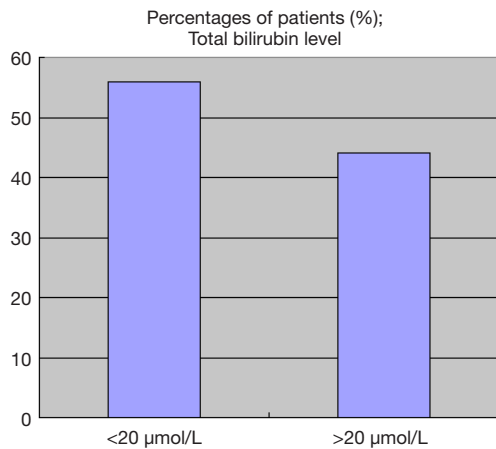
A total of 40 children (male: 24; female: 16) were included in the study. Among them, 25 children had good compliance and were followed up at least twice. The patients’ ages at the last follow-up were  $13.38 \pm 21.70$  months. The age distribution is shown in *Figure 1*. A total of 27 children were 0–1 years old, 5 children were 1–3 years old, 7 children were 3–6 years old, and 1 child was 6–9 years old. Nineteen children were followed up only once; the main reasons for the loss to follow-up were decreased treatment intention and waiting for a liver source in another city.

**Pre- and post-KPE liver function**

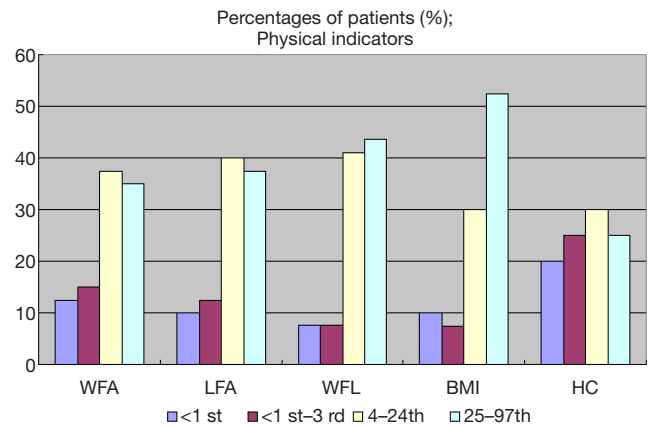
The pre- and post-KPE regular monitoring of liver function were compared (*Table 1*). The aspartate aminotransferase (AST), total protein (TP), globulin (GLO), albumin to globulin ratio (A/G), TBIL and direct bilirubin (DBIL) were all significantly improved after KPE.

**Table 1** Pre- and post-KPE liver function

Liver function index	N	Pre-KPE	N	6 months post-KPE	T value	P value
ALT (U/L)	16	189.00±107.51	25	131.36±91.94	1.8330	0.0744
AST (U/L)	16	235.38±125.42	25	128.76±87.80	3.2054	0.0027
ALP (U/L)	15	451.13±215.55	23	389.78±154.06	1.0243	0.3125
Gamma-GT (U/L)	16	944.50±551.23	24	989.25±2,031.62	-0.0857	0.9322
TP (g/L)	16	57.93±5.00	25	64.36±6.37	-3.4166	0.0015
ALB (g/L)	16	39.06±3.52	25	40.67±4.76	-1.1672	0.2502
GLO (g/L)	16	18.87±4.84	25	23.68±4.38	-3.2981	0.0021
A/G	16	2.18±0.49	25	1.78±0.41	2.8141	0.0076
TBIL (µmol/L)	16	123.60±30.72	25	62.54±86.15	2.7159	0.0098
DBIL (µmol/L)	16	102.28±24.84	25	48.86±70.73	2.8979	0.0061
IBIL (µmol/L)	16	21.32±8.35	25	13.69±15.80	1.7740	0.0839
TBA (µmol/L)	15	168.19±92.17	25	128.47±134.85	1.0058	0.3209
PA (mg/L)	9	130.83±70.56	20	150.82±47.48	-0.6437	0.5268



**Figure 2** The jaundice clearance rate within 3 months after KPE.



**Figure 3** The percentile distributions of individual physical indicators at the last follow-up.

**Table 2** The physical statuses of the children at different periods

Physical index	Birth	Admission	Last follow-up
Gestational age/ month	38.65±2.00	1.91±1.04	13.38±21.7
Measured value			
Weight (kg)	3.12±0.54	4.9±0.78	6.54±2.46
Length (cm)			67.01±10.63
HC (cm)			41.24±4.12
BMI			15.65±2.11
Z-score			
Weight	-0.34±1.02	-0.51±1.09	-1.49±1.54
Length	0.14±1.18		-1.39±1.16
WFL	-0.82±1.35		-0.48±1.13
BMI			-0.65±1.29
HC			-1.43±1.93
Percentile			
Weight	40.57±28.9	33.17±27.48	26.53±25.91
Length	54.2±31.83		18.93±18.96
WFL	35.32±30.45		37.95±30.69
BMI			37.5±31.36
HC			22.72±26.52

WFL, weight for length; HC, head circumference; BMI, body mass index.

**Jaundice clearance rate within three months after KPE**

The post-KPE TBIL level is shown in *Figure 2*. Twenty-five children were followed up with blood sampling, and the TBIL of 14 children (14/25, 56%) decreased to less than 20 μmol/L.

**The pre- and post-KPE physical statuses**

The pre- and post-KPE physical statuses are shown in *Table 2*.

The percentile distributions of individual physical indicators at the last follow-up visit are shown in *Figure 3*. The incidence rate of malnutrition was as follows. In the weight for age (WFA) comparison, 11 children’s weights were less than the 3rd percentile (11/40, 27.5%), and 5 were less than the 1st percentile (5/40, 12.5%). In the length for age (LFA) comparison, 9 children were less than the 3rd percentile (9/40, 22.5%), and 4 were less than the 1st percentile (4/40, 10%). When the weight for length (WFL) was compared to the reference standards, 6 of the children’s WFL was less than the 3rd percentile (6/39, 15.4%), and 3 were less than the 1st percentile (3/39, 7.7%). In the comparison of their BMIs with those of their peers, 7 children’s BMIs were less than the 3rd percentile (7/40, 17.5%), and 4 were less than the 1st percentile (4/40, 10%). Compared with the head circumference (HC) of their peers, 18 children’s HCs were less than the 3rd percentile (18/40, 45%), and 8 were less than the 1st

**Table 3** Fat-soluble vitamin levels after KPE

Fat-soluble vitamin	Post-KPE group (n=22)	Control group (n=30)	T value	P value
Vitamin A ( $\mu\text{mol/L}$ )	0.99 $\pm$ 0.29	0.94 $\pm$ 0.12	0.425	0.674
Vitamin D ( $\mu\text{mol/L}$ )	34.12 $\pm$ 16.71	40.00 $\pm$ 17.80	-0.754	0.458
Vitamin E ( $\mu\text{g/mL}$ )	12.82 $\pm$ 1.53	11.61 $\pm$ 0.47	0.423	0.676

percentile (8/40, 20%).

### *Post-KPE fat-soluble vitamin levels*

The fat-soluble vitamin levels in children with a jaundice clearance rate of 56% after KPE are shown in *Table 3*. BA patients are a high-risk group for vitamin deficiency. After supplementation with the physiological dose of vitamin D (400–800 IU), their fat-soluble vitamin levels were not significantly different from those of children of the same ages from the outpatient clinics.

## Discussion

For every 2,500 full-term infants, one child is born with cholestatic jaundice. The most common cause of neonatal cholestatic jaundice for newborn babies is BA (25–40%), which includes single-gene genetic disorders (25%) and other unknown or multifactorial causes (such as intravenous nutrition); each of these cases has its own time sensitivity and treatment plans (2). BA is an infantile disease with progressive fibrosis and obstruction of the intrahepatic and extrahepatic bile ducts. Its incidence rate varies slightly worldwide, with 1/19,000 among live born infants in Europe (10) and 1/18,000 among live born infants in North America (11). Children with this disease eventually develop end-stage liver disease requiring LT (6). The standard BA surgical procedure is KPE. Early diagnosis is extremely important, especially in infants with jaundice for up to 6 weeks. An earlier diagnosis leads to more successful KPE. Intraoperative cholangiogram evaluation and histological examination of the residual biliary duct are the gold standard for diagnosis of BA (2). Different from other cholestatic liver diseases, certain characteristics of the liver histopathology in BA patients are independent factors predicting high-risk LT, such as advanced-stage fibrosis, pancreatic duct plate structure, medium and advanced degree of bile duct damage, relatively old age when KPE is performed and elevation of the international normalized ratio (INR) (12).

Although nearly 50% of these children may need an LT before 2 years old, the other 50% may live with autologous livers for years without cirrhosis or chronic liver disease. However, many children suffer from a variety of chronic complications, such as recurrent cholangitis, portal hypertension, bleeding from varicose veins, growth problems, biochemical abnormalities and bone malnutrition. The incidence of these complications affects the quality of life of the children and their families (13).

Although nearly 50% of the children survive with autologous livers for more than 10 years after KPE (13,14), a 20-year study performed in Northern European countries showed that the treatment survival intention of BA children was lower than that of other children with chronic liver disease waiting for LT. Because these children have a younger age (<1 year), lower weight (<10 kg), higher bilirubin (>510  $\mu\text{mol/L}$ ), higher INR (>1.6), and higher pediatric end-stage liver disease (PELD) score (>20), they have the worst outcome (15). Children younger than 2 years of age with end-stage liver disease (YC2) are a uniquely vulnerable group awaiting LT, most of whom are children with BA. Multivariate analysis suggested that for YC2 waiting for LT, a PELD >21, body length <60.6 cm, weight >10 kg and initial creatinine >0.5 were independent risk factors of mortality (16). This finding suggests that the nutritional status after KPE and before LT is very important for the survival of BA patients.

KPE was performed on 106 children with type III BA who were divided into 2 groups according to the STRONGkids score: moderate nutritional risk (MNR) and high nutritional risk (HNR). The children in the HNR group were more prone to cholangitis, the post-KPE jaundice clearance rate for these children was lower than that in the MNR group, and the occurrence of the first postoperative cholangitis in the MNR group was later than that in the HNR group. Two-year survival was also significantly higher in the MNR group than in the HNR group (5). Therefore, nutritional support should be timely and effective. Although drugs such as steroid therapy can improve the short-term ( $\leq 1$  year) jaundice clearance rate



after KPE, no significant improvement has been noted in the long-term ( $\geq 2$  years) jaundice clearance rate and the survival rate of children with autologous livers (17). The early nutritional status after KPE was found to be a predictive factor for the survival of children with autologous livers at the age of 2 years. The TBIL and albumin levels 3 months after KPE were independently correlated with the survival rate of children with autologous livers based on data from 217 BA children. Even in patients whose jaundice is completely cleared after KPE, an albumin level below 35 g/L indicates a poor prognosis (18). Therefore, improving the nutritional status after KPE is extremely important.

In this study, a nutritional follow-up visit was conducted with 40 children with a jaundice clearance rate up to 56% after KPE. We found that nutritional disorders appeared in the early stage. Regarding the incidence of malnutrition, 27.5% of the children had a weight lower than that of their peers, 15.4–17.5% were emaciated, 22.5% had a short stature, and 45% had a smaller-than-normal head circumference. These results differed from those of Mansi *et al.* (19), who investigated hospitalized children with chronic liver disease and found that 35.6% had a low weight, 49% had a short stature, and 5–10% were emaciated. In terms of the types of malnutrition, short stature is more concerning, because it reflects the long-term nutritional status of children, whereas weight reflects the short-term nutritional status. A total of 2/3 of these children are under the age of 1 year, nearly 80% of them are younger than 3 years old, and the percentage with a short stature is significantly higher than the percentage who are emaciated. Although KPE was completed as early as 2–3 months of age and obstructive jaundice was alleviated in these children, some of them still suffered from significant impairment of the linear growth rate.

In the comparisons of bilirubin, liver function and albumin before and after KPE, the efficacy of KPE for these children was significant, because TBIL, DBIL and AST decreased significantly, total protein increased significantly, and the jaundice clearance rate reached 56% within 3 months after KPE. If jaundice clearance is achieved within 3 months after KPE, significant improvement in the nutritional status of the patients can be expected. Studies have shown that children with BA are prone to deficiency of fat-soluble vitamins, especially vitamin D (7). Therefore, a vitamin D formulation should be routinely supplemented. In this study, nutritional interventions were conducted in a timely manner for children at high nutritional risk. At the follow-up visit, no significant differences in the fat-soluble vitamin levels were found compared with those of children

of the same ages from the outpatient clinic. The liver is an important metabolic organ that plays a major role in integrating various biochemical pathways for metabolism. Therefore, the long-term nutritional and growth statuses still need to be followed up. In addition to supplementation with fat-soluble vitamins and the MCT-enriched nutritional formulation implemented in this study, we suggest that clinical dieticians be included in the multidisciplinary team (MDT) that will follow-up the patients after KPE to develop more comprehensive nutritional treatment and follow-up strategies, such as more sophisticated management of the relationship between drugs, foods and nutritional supplements and the sequence of administration.

In conclusion, long-term follow-up should be conducted for BA patients after KPE, and their growth and nutrient levels need to be monitored dynamically to improve the long-term survival rate of the children and particularly to strive for an optimal nutritional level before LT (10,20). Our study has some shortcomings: The follow-up time is relatively short, the rate of loss to follow-up is relatively high, and the number of cases is relatively few. In the future, we hope to standardize the follow-up system, increase the number of cases, group the children based on degrees of jaundice clearance after KPE, and perform a more accurate nutritional assessment to provide an objective basis for individualized nutrition management.

## Conclusions

Among the children with BA who survived with autologous livers after KPE, 56% (14/25) achieved jaundice clearance 3 months after KPE. At the last follow-up at 13.38 $\pm$ 21.70 months, 32.5% (13/40) of the children were in the ideal weight range of P25–P75, 37.5% (15/40) were in the ideal body length/height range, 28.2% (11/39) were in the ideal WFL range, and 42.5% (17/40) was in the ideal BMI range.

At the last follow-up, the emaciation rate was 17.5% (7/40), and the short stature rate was 22.5% (9/40), indicating that long-term malnutrition was prominent.

Long-term standardized follow-up and nutrition management for children after KPE should be carried out with multidisciplinary cooperation to achieve a more ideal nutritional level before LT.

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## Footnote

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*Conflicts of Interest:* All authors have completed the ICMJE uniform disclosure form (available at <http://dx.doi.org/10.21037/dmr.2019.07.03>). The series “Nutritional Support for Digestive Surgery” was commissioned by the editorial office without any funding or sponsorship. The authors have no other conflicts of interest to declare.

*Ethical Statement:* The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the ethics committee of The Guangzhou Women and Children Medical Center Research Ethics Committee (No. 26001). Informed consent was obtained.

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