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Original Article

Comparison of nutritional status among children with biliary atresia according to age at the time of Kasai procedure

Febricilla Citra Pratiwi, Titis Widowati, Endy P. Prawirohartono

Abstract

Background Recent studies revealed better outcomes among children with biliary atresia (BA) who underwent Kasai procedure at 60 to 90 days of age. Delayed Kasai procedure has a high risk of complications, including nutritional deficits which lead to malnutrition.

Objective To determine the nutritional status of children with BA according to age at the time of Kasai procedure.

Methods Using medical records, we conducted a retrospective study in children with BA based on magnetic resonance cholangiopancreatography (MRCP) or intraoperative cholangiography, who were admitted between 2015 to 2017 and underwent Kasai procedure at Dr. Sardjito General Hospital, Yogyakarta. Nutritional status was evaluated on the day before Kasai procedure, and classified into 4 groups of subjects based on age at the time the children underwent Kasai procedure (<60 days, 60-90 days, >90-120 days, and >120 days). Normal distribution data was analyzed with Saphiro-Wilk test and mean T-test was used to compare mean age at the time of Kasai procedure between groups of well-nourished and malnourished subjects.

Results A total of 39 children with BA underwent Kasai procedure. Of 3 children who underwent Kasai procedure at <60 days of age, 2 were well-nourished and 1 was malnourished. Of the 12 children who underwent Kasai procedure at 60-90 days of age, 6 were well-nourished and 6 were malnourished. Of the 7 children who underwent Kasai procedure at >90-120 days of age, 4 were well-nourished and 3 were malnourished. Of 17 children who underwent Kasai procedure at >120 days of age, 5 were well-nourished and 12 were malnourished. The means of age at the time of Kasai procedure were higher in malnourished subject than well-nourished.

Conclusion The highest prevalence of malnourishment is seen in children with biliary atresia who underwent Kasai procedure at >120 days of age. **[Paediatr Indones. 2019;59:294-7; doi: http://dx.doi.org/10.14238/pi59.6.2019.294-7]**.

Keywords: *biliary atresia; children; nutritional status; Kasai procedure*

Biliary atresia is an obstructive cholangiopathic disease of unknown etiology, characterized by inflammatory destruction of the intra- and extrahepatic bile ducts, which causes bile flow obstruction and leads to cholestasis and cirrhosis.[1,2] The current management of biliary atresia patients is to relieve the extrahepatic biliary obstruction and restore bile flow by performing Kasai procedure while waiting for a liver transplant.[3]

Age of diagnosis plays a major role on biliary atresia management. Delayed Kasai surgery impacts bile retention leading to many severe complications.[3] Malnutrition is the most common complication in untreated biliary atresia patients. It frequently occurs within the first few months of life caused by nutritional deficits. The earlier the malnutrition occurs, the higher risk of a poor prognosis. Therefore,

From the Department of Child Health, Faculty of Medicine, Public Health and Nursing, Gadjah Mada University/Dr. Sardjito Hospital, Yogyakarta, Indonesia.

Corresponding author: Febricilla Citra Pratiwi, Department of Child Health/Faculty of Medicine, Public Health and Nursing, Gadjah Mada University/Dr. Sardjito General Hospital, Jl. Kesehatan No. 1, Sekip Utara, Yogyakarta 55281, Indonesia. Tel. +62-274-489726/+62-274-561616; Fax. +62-274-583745; Email: febricillacitra@yahoo.com.

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the aim of our study was to evaluate the pre-operative nutritional status of biliary atresia patients according to the age at which they underwent Kasai procedure.

Methods

We conducted a retrospective study among children with biliary atresia based on magnetic resonance cholangiopancreatography (MRCP) or intraoperative cholangiography who were admitted between January 2015 and December 2017 and underwent Kasai procedure at Dr. Sardjito General Hospital, Yogyakarta. Data were collected from patients' medical records using a questionnaire covering demographic data (age and sex) as well as clinical data which included date of admisson, date of diagnosis, date of Kasai procedure, type of biliary atresia, weight, and height on the day before Kasai procedure.

Nutritional status was evaluated on the day before Kasai procedure and subjects were classified into well-nourished or malnourished, using weight-for-height *z*-scores. Malnourished status was defined as having a weight-for-height *z*-score of less than -3.0 to less than -2.0 based on the WHO Child Growth Standard 2006.[4] Patients with incomplete medical records were excluded from the study. Subjects were divided into four age groups according to their age at the time of Kasai procedure: <60 days, 60-90 days, >90-120 days, and >120 days. Data were analyzed using *SPSS version 20.0* software (*SPSS Inc*., Chicago, IL, USA). The data of subjects were tested for normal distribution using the Saphiro-Wilk test before calculation of differences. Univariate analysis was conducted to describe the characteristics and distribution of research data. Mean T-test was used to compare mean age at the time of Kasai procedure between groups of well-nourished and malnourished subjects.

A P-value of <0.05 was consindered statistically significance. Results were presented in the form of descriptive narrative and tables. This study received approval from the Medical and Health Research Ethics Committee of the Faculty of Medicine, Public Health and Nursing, Gadjah Mada University/ Dr. Sardjito General Hospital, Yogyakarta, Indonesia.

Results

A total of 39 children who had been diagnosed with biliary atresia and underwent Kasai procedure at Dr. Sardjito General Hospital, Yogyakarta between January 2015 and December 2017 were included in the study. Characteristics of subjects are shown in **Table 1**. Of our subjects, 44% were well-nourished and 56% were malnourished.

Table 1. Basic characteristics of study subjects

Characteristics	N =39
Gender, n	
Male	22
Female	17
Median age of admission (IQR), day	103 (15 - 323)
Median age of Kasai procedure (IQR), day	118 (46 – 324)
Type of biliary atresia, n	
Type 1	2
Type 2	16
Type 3	18
Unclassified	15
Age at Kasai procedure, n	
<60 days	3
60-90 days	12
>90-120 days	7
>120 days	17

Subjects' nutritional status according to age at the time of Kasai procedure is shown in **Figure 1**. These children underwent Kasai procedure at <60 days of ages, 2 out of 3 were well-nourished. Of the 12 patients who underwent Kasai procedure at 60-90 days of age, 6 were well-nourished and 6 were malnourished. Of the 7 patients who underwent Kasai procedure at >90-120 days of age, 4 were well-nourished and 3 were malnourished. Of 17 patients who underwent Kasai procedure at >120 days of age, 5 were well-nourished and 12 were malnourished. The highest proportion of malnourished subjects was in the >120 days of age group.

The means of age at the time of Kasai procedure were significantly higher in malnourished subject than well-nourished (**Table 2**).

Table 2. Univariate analysis of nutritional status according to age at the time of Kasai procedure

Variables	Mean age at the time of Kasai procedure (SD), days	P value
Nutritional status		0.021
Well-nourished	101 (33)	
Malnourished	137 (59)	

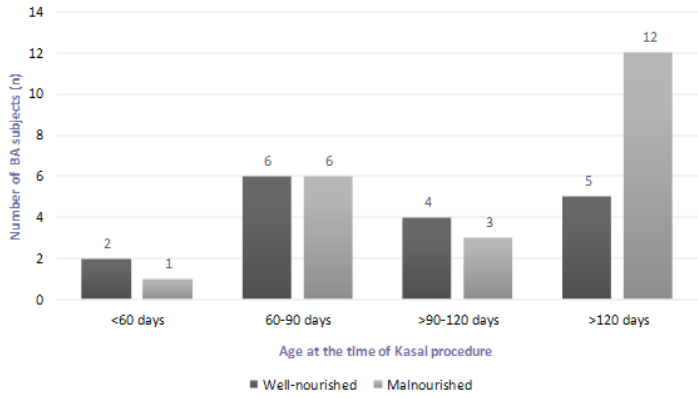

Figure 1. Proportion of nutritional status in BA patients according to age at the time of Kasai procedure

Discussion

The median age of children with biliary atresia who underwent Kasai procedure in our hospital was 118 days, and 92% of subjects underwent Kasai surgery after 60 days of life. Previous studies have confirmed that timing of management of children with biliary atresia, including diagnosis and Kasai procedure, had an important role and was directly correlated to prognosis.[1,3] Children with biliary atresia who undergo Kasai procedure before 60 days of age have a high likelihood of obtaining good initial bile flow (80-90%), improved liver funtion, and fewer complications.[1,2] Increased age at Kasai procedure led to a progressive and lasting detrimental effect on outcome. Obstruction of the biliary tract between the liver and intestines results in bile retention and causes destruction of the liver. Thus, performing the Kasai procedure in older infants reduces its success because of hepatic cirrhosis and greater complications.[3]

Malnutrition is a common complication in biliary atresia patients. Several studies have provided evidence that malnutrition was associated with prognosis in patients with liver disease. Malnourished patients exhibited a significantly increased incidence of post-operative infection, delayed recovery from illness, and increased mortality after surgery.[5] In our study, 56% of biliary atresia patients were malnourished before Kasai procedure, similar to previous studies which reported proportions of 46-53%.[1,5] Biliary atresia patients tend to present with nutritional deficiencies caused by multiple factors, including increased energy and nutrient loss, reduced calorie intake, alteration of nutrient metabolism, and greater nutritional needs. Due to inadequate nutrient intake and increased consumption, biliary atresia patients frequently exhibit protein-energy malnutrition.[5] Children with biliary atresia have poor bile flow, resulting in reduced delivery of bile acids to the small intestine, which impairs micelle formation necessary for absorption of fat-soluble vitamins A, D, E, and K, as well as subsequent fat and fat-soluble vitamin malabsorption.[1,6] In addition, children with biliary atresia have poor appetites and higher resting energy expenditure than healthy normal children.[1]

Malnourishment was found in every age group (**Figure 1**), with greater numbers in the oldest age group. Most of our subjects who underwent Kasai procedure at more than 120 days of age presented with malnutrition, which was evaluated on the day before Kasai procedure. Our results indicate that delaying Kasai procedure to >120 days of age contributes to higher risk of malnutrition complications.

Reasons for delayed Kasai procedure include late recognition and/or referral of biliary atresia patients. Delayed recognition of biliary atresia is caused by the difficulties in differentiating jaundice symptoms from other jaundice disorders and identifying abnormal stool color. A lack of awareness in primary health care workers or physicians for referring children with persistent jaundice for specialty care is also a major problem in most parts of the world, especially in developing countries, including Indonesia. A previous study noted that in India, only 20% cases that presented in most health centers were aged less than 60 days.[7]

A limitation of this study was that we cannot show a causal relationship between variables. With regards to higher prevalence of malnutrition in the older age group, we recommend improving the awareness and ability of our primary healthcare workers or physicians to immediately identify and refer infants with persistent jaundice, as well as to consider the critical importance of nutritional status assessment. Attention should also be paid to metabolic changes in biliary atresia patients. The appropriate nutritional support may reduce the incidence of malnutrition and improve the outcome.

In conclusion, malnourishment in children with BA is higher in those who undergo Kasai surgery at an older age. We note the highest prevalence of malnourishment in biliary atresia patients who underwent Kasai procedure at >120 days of age. Kasai procedure is recommended within 60-120 days of life to attain good nutritional status.

Conflict of interest

None declared.

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