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Quality of Life of Parents and Patients with Biliary Atresia in Native Liver

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ABSTRACT

Children with chronic conditions such as biliary atresia may experience several issues that affect their quality of life (QoL). Childhood chronic disease impacts family members and leads to family health impairment. Objective**:** This study aims to assess the QoL of parents and patients with biliary atresia. Methods and material: The assessment of parent and patient quality of life with biliary atresia was conducted using the Pediatric Quality of Life (PedsQL) parent report and the World Health Organization Quality of Life (WHOQoL). Multivariate linear regression was used for associated factors. This case-control study involved 38 children (0-24 months) with biliary atresia at the Dr. Soetomo General Academic Hospital Surabaya, which was conducted from June to July 2023. Data obtained showed that male and female prevalence was equal (50%). Age <6 months, mothers with high school education, family income >3 million rupiahs, and normal nutritional status were frequently in patients with biliary atresia. Based on the WHOQoL questionnaire, the environment was the only domain that impacted the quality of life of the parents significantly (P= 0.005). However, no significant association with patients' QoL was found for the five domains of the PedsQL. The environmental factor had a significant impact on the QoL of the patient's parents according to the results of the WHOQoL questionnaire (P= 0.005).

KEYWORDS: Biliary Atresia, PedsQL, WHOQOL, Quality of Life, Children .

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INTRODUCTION

Biliary atresia is a progressive fibroinflammatory disease of bile ducts. It is a major cause leading to liver transplantation in children (Rohani *et al.*, 2022). Biliary atresia is characterized by the presence of jaundice, hepatomegaly, and acholic stools (Palladini, 2015). Early diagnosis of BA is crucial because early intervention with Kasai procedure has best outcomes and prevents liver transplantation (Le *et al*., 2022).

Children with chronic disease can impact family members and have a negative effect on family health (Golics *et al*., 2013). Parents of children with chronic disease have a higher level of stress associated with their role as caregivers. Furthermore, the stress related to the parent-child relationship also increased. The increase in parental stress is primarily linked to health-related factors (Pinquart, 2018). The combination of trait anxiety and alterations in the parental role explained 26% of the variability in the maternal state of anxiety (Lisanti *et al*., 2017).

Quality of Life (QoL), according to the WHOQoL, is a subjective evaluation of their overall well-being, taking into account their social and cultural background, personal aspirations, and concerns (Group *et al.*, 1994). The health-related quality of life (HRQoL) of children who receive a liver transplant has been shown to be reduced compared to healthy controls, although their HRQoL is similar to those of other illness groups and solid organ recipients (Parmar et al., 2017). The HRQOL in biliary atresia with native liver is worse than in healthy children and similar to children with biliary atresia who undergoing liver transplantation (Sundaram *et al.*, 2013). The physical function scores were found to be higher in biliary atresia patients with adequate health status compared to those who had inadequate health condition (Feldman *et al.*, 2016).

This study aimed to assess the quality of life, levels of anxiety, and levels of stress experienced by parents of children with biliary atresia and compare with healthy children.

RESEARCH METHODS

A case-control HRQoL study was conducted in children with biliary atresia aged 0-24 months. We identified eligible children diagnosed with biliary atresia who visited the Pediatric Hepatology Outpatient Clinic. The sample was divided into two groups, healthy children and biliary atresia children. The assessment of quality of life in parents and patients with biliary atresia using of the PedsQL and WHOQoL measurements. The inclusion criteria for this study was children with biliary atresia aged 0-24 months. The exclusion criteria was insufficient knowledge to fill the questionnaire or no inform consent obtained from the parents. Ethical clearance was obtained from the Soetomo Hospital Ethics Committee.

Patients that came to Pediatric Hepatology Outpatient Clinic with biliary atresia that meet the inclusion criteria was given the inform consent. Following acquiring informed consent, parents were requested to complete the forms. The questionnaire consists of demographic of the patients including age, gender, perinatal history as well as information about the parents, such as parental age, educational status, and income. In order to evaluate parental QoL we used the WHOQoL questionnaires which consist of 26 items assessing four domains which are physical health, mental health, social interactions, and environment. A higher domain scores shows an improved quality of life. We used the parent report PedsQL to assess the patients' QoL which contained 5 domains: physical function, physical symptoms, emotional function, social function, and cognitive function. Scoring for parent report PedsQL was concluded into two groups: psychosocial health summary score and physical health summary score. The psychosocial health summary score is calculated by taking the average of the emotional function, social function, and cognitive function meanwhile the physical health summary score is calculated by taking the average of the emotional function, social function, and cognitive function is mean of the physical function and physical symptoms. We compared parents' and patients' QoL of healthy children vs children with biliary atresia.

We presented participant characteristics as either mean or median range depending on the data distribution. Regarding the QoL, higher scores shows better QoL. We used Pearson's correlation test to compare outcomes between subgroups. A p-value <0.05 was considered as significant Utilizing IBM SPSS version 23 for the purpose of data analysis.

RESULTS

The case-control study included a total of 38 children (0-24 months) with biliary atresia (n= 18) and health control (n=20).

Table 1. Characteristics of case and control group

Parameter	Case (n=18)	Control (n=20)	P value
Patient's Age (months)	7 (4.75 ; 43.25)	8.5 (5.25 ; 9.75)	0.649
Maternal Age (years)	28 (± 6.58)	30 (± 3.25)	0.176
Paternal Age (years)	32,94 (± 5.92	32.35 (± 3.45)	0.700
Sex			
Male	9	9	0.758
Female	9	11	0.758
Perinatal History			
Aterm	18	17	0.232
Preterm	0	3	0.232
Vaginal births	9	5	0.11
Cesarean births	9	15	0.11

Table 1 showed characteristic in both case and control group. The data obtained showed that in children with biliary atresia the prevalence of boys and girls was similar (50%).

Among patients with biliary atresia, age <6 months, mothers with high school education, and family income >3 million rupiah were more frequent (Table 2).

Table 2. Characteristics of children with biliary atresia

Parameter	%
Age (months)	
< 6 months	66%
> 6 months	44%
Sex	
Male	50%
Female	50%
Mothers Education	
Primary School	6%

High School	72%
Diploma	16%
Bachelor	6%
Family Income	
< 1.000.000	28%
1.000.000-3.000.000	22%
> 3.000.000	50%

Table 3. Patients' and parents' QoL

Outcome	Mean (Range)		p-value
	Case	Control	
Parent Report PedsQL			
Psychosocial Health Summary Score	74.3 (65.6-83.1)	82.5 (70.6-93.1)	0.077
Physical Health Summary Score	84.3 (72.6-93.7)	92.1 (80-98.4)	0.155
WHOQoL			
Physical Health	69 (56-76.5)	69 (63-78)	0.719
Psychological	69 (56-81)	69 (57.7-69)	0.394
Social Relationship	69 (54.5-75)	75 (59.2-75)	0.163
Environment	63 (54.5-69)	72 (63-75)	0.049

The higher the score of quality of life indicates a better quality of life. Psychosocial health summary score (median = 74.3, p-value = 0.077) and physical health summary score (median = 84.3, p-value = 0.155) in children with biliary atresia have a comparable score compared to healthy children. According to the WHO QoL questionnaire, the environment was the only domain which associated with parental QoL (p-value = 0.005).

DISSCUSSION

Biliary atresia is fibroinflammatory of the intrahepatic and extrahepatic bile ducts. Biliary atresia causes prolonged neonatal jaundice and pale stools (Lakshminarayanan and Davenport, 2016). Kasai surgery is currently used to restore bile drainage, however outcomes are variable and liver transplantation may be required (Bezerra *et al*., 2018). Children with biliary atresia are at risk for progressive liver disease, with persistent cirrhosis even after biliary atresia surgery (Hussain *et al.*, 2017; Chan *et al.*, 2019). Complications of biliary atresia, such as recurrent cholangitis and portal hypertension, increase morbidity and mortality (Gad *et al*., 2021; Ng *et al*., 2014). Quality of life (QoL) assessment in biliary atresia has increasingly received attention due to improved survival with evolving therapeutic modalities. QoL evaluation includes the effect on physical and mental health as a result of chronic disease (Hukkinen et al., 2022).

Parents of children with severe liver disease leading to liver transplants suffer dysfunctional family roles and psychological problems (Zhang *et al*., 2023). Previous studies showed that there was no major disparity in QoL between parents of children with a native liver and those who received liver transplantation. Physical quality of life is inversely correlated with older age and high levels of parental concern. However, lower income family and high level anxiety were found negatively correlation with environmental QoL (Rodijk *et al.*, 2020). Parental psychological status and coping strategies have an impact on the physical and mental health of newborns (Zhang *et al*., 2023). Studies show that parents, especially mothers, report high anxiety when dealing with a child diagnosed with biliary atresia. High levels of anxiety and stress have a significant correlation with lower quality of life for both mothers and fathers (Rodijk *et al*., 2022).

Compared to healthy children, children with biliary atresia who received a transplant and their parents had lower physical function scores (Feldman *et al*., 2016). Female gender, higher total bilirubin and number of immunosuppressives were associated with worse HR QoL. On the other hand, older age at the time of the survey was associated with better HR QoL in biliary atresia.(Le *et al*., 2022; Miserachs *et al.*, 2019). Children with biliary atresia living with a native liver had a lower general and physical health score significantly (Wong *et al*., 2018).

CONCLUSIONS

Providing high-quality healthcare for children with biliary atresia and their families requires the combination of evidence-based clinical care and the promotion of a positive experience.

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