

**Biliary atresia and its impacts on the affected child and his family:
an integrative review**

**Atresia biliar e seus impactos para a criança acometida e sua família:
revisão integrativa**

**La atresia biliar y sus repercusiones en el niño afectado y su familia:
revisión integradora**

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Abstract: Objective: To identify through literature the impacts that biliary atresia causes in affected children and their families. Method: This is an integrative review of literature carried out between July and September 2022, using databases and libraries: SCOPUS, PUBMED, and EBSCO. The descriptors for the search were adjusted according to the database or library, being caregivers, “biliary atresia”, “child care”, family, child, “child care subject”, combined with the boolean operator AND. The studies were selected in a careful and systematic way and evaluated for the level of evidence. Results: In the primary search, 1441 articles were identified, and after careful selection, 7 studies were selected to compose the qualitative analysis. The main results of the impacts of biliary atresia for children refer to the consequences arising from the disease regarding the need for surgical procedures that negatively impact quality of life. The impacts for the family relate to consequences for mental health and low quality of life, combined with lack of knowledge about caring for the child and excessive worry. Conclusion: This study made it possible to identify the impacts that biliary atresia causes in affected children and their families, pointing to significant contributions to nursing care.

Keywords: biliary atresia; caregivers; family health; child health.

Resumo: Objetivo: Identificar através da literatura impactos que a atresia biliar provoca em crianças acometidas e em sua família. Método: Trata-se de uma revisão integrativa da literatura realizada entre julho e setembro de 2022, utilizando bases de dados e bibliotecas SCOPUS, PUBMED e EBSCO. Realizou-se adequação dos descritores para busca de acordo com a base ou biblioteca, sendo estes: caregivers, “biliary atresia”, “child care”, family, child, “child care subject”, combinados com o operador booleano AND. Os estudos foram selecionados de forma criteriosa e sistematizada, avaliados quanto ao nível de evidência. Resultados: Na busca primária, identificou-se 1441 artigos e após criteriosa seleção, 7 estudos foram selecionados para compor a análise qualitativa. Obteve-se como principais

resultados dos impactos da atresia biliar para as crianças, dizem respeito as consequências oriundas da doença no que se refere a necessidade de procedimento cirúrgico que impactam negativamente na qualidade de vida. Já os impactos para a família dizem respeito a consequências para a saúde mental e baixa qualidade de vida aliados ao desconhecimento sobre cuidados com a criança e excesso de preocupação. Conclusão: Este estudo possibilitou a identificação dos impactos que a atresia biliar provoca em crianças acometidas e em sua família, apontando contribuições significativas para o cuidado em enfermagem.

Palavras-chave: atresia biliar; cuidadores; saúde da família; saúde da criança.

Resumen: Objetivo: Identificar a través de la literatura los impactos que la atresia biliar provoca en los niños afectados y en su familia. Método: Se trata de una revisión integradora de la literatura, realizada entre julio y septiembre de 2022, utilizando como bases de datos y bibliotecas SCOPUS, PUBMED y EBSCO. Se realizó la adecuación de los descriptores en busca de concordancia con la base o biblioteca, siendo estos: caregivers, “biliary atresia”, “child care”, family, child, “child care subject”, combinados con el operador booleano AND. Los estudios fueron seleccionados de forma juiciosa y sistematizada, evaluados en cuanto al nivel de evidencia. Resultados: En la búsqueda primaria fueron identificados 1441 artículos y, después de una cuidadosa selección, 7 estudios fueron seleccionados para componer el análisis cualitativo. Los principales resultados de los impactos de la atresia biliar para los niños son las consecuencias de la enfermedad con relación a la necesidad de cirugía, que impactan negativamente en la calidad de vida. Por otro lado, los impactos para la familia están relacionados a las consecuencias para la salud mental y baja calidad de vida, asociados a la falta de conocimiento sobre el cuidado del niño y preocupación excesiva. Conclusión: Este estudio permitió identificar los impactos que la atresia biliar provoca en los niños afectados y su familia, y aporta contribuciones significativas para el cuidado en enfermería.

Palabras claves: atresia biliar; cuidadores; salud de la familia; salud infantil.

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Introduction

Biliary atresia is an obstructive and progressive process of unknown etiology that affects the intrahepatic or extrahepatic bile ducts in newborns and young infants and causes severe cholestatic jaundice and cirrhosis. ⁽¹⁾ Its anatomical classification is divided into: i) Type I atresia, which occurs in 5-12 % of cases, with common bile duct obliteration, with free transit through proximal canals; ii) Type II atresia, which occurs in 2 % of cases, and may be common hepatic duct atresia, with or without common bile duct, cystic and gallbladder atresia or common bile duct atresia, cystic atresia and gallbladder atresia with

patent common hepatic duct; and iii) Type III atresia, which occurs in 86-90 % of cases, with atresia of the common bile duct, liver and gallbladder. ⁽²⁾

Biliary atresia is the main cause of liver transplantation in children. ⁽³⁾ The early disease diagnosis is indispensable, because when the surgical treatment —Kasai portoenterostomy— is performed up to 60 days of life, the restoration of bile flow is obtained in up to 70 % of patients and in those operated with more than 90 days of life, in less than 25 %. However, it should be noted the late referral of children with Biliary Atresia is still a sad reality in Brazil and in the world. ⁽⁴⁾

While confronted with a disease that requires such complex therapy, it is important to reflect on the concerns parents/caregivers develop for the child. Seemingly child hospitalization generates suffering and trauma, both for the child, who experiences atypical experiences of its routine, and for the parents, due to anxieties both in relation to the presence of the child's suffering and in relation to the therapeutic plan. ⁽³⁾

When it comes specifically to parents whose children have developed biliary atresia, studies indicate about 85 % of parents report extreme concern when hearing the diagnosis of biliary atresia. It is noteworthy over time and among children who developed good health after diagnosis, parents' concern decreased. ⁽¹⁻⁵⁾ In addition, considering biliary atresia is the main cause of childhood liver transplantation, it is worth noting this procedure has a direct impact on the mental health of parents and this has a direct impact on the care of children with biliary atresia. ⁽¹⁾

In addition to concerns about the sick child, parents/caregivers should also address concerns related to post-transplant care. Some general care in the post-transplant period concerns care with food and water intake, cleaning, administration of medications and emotional support to the child. ⁽⁶⁾

As mentioned before, this study aimed to identify through the literature impacts biliary atresia causes in affected children and their families. This study became important because it was possible to summarize evidence corroborates for teaching, research and care to the child and family caregiver beyond care with the disease itself, but also encompassing other consequences the problem causes.

In teaching, parental health education is highlighted with regard to the care of children in the aforementioned situation, which can impact on reduction of distress and complications' prevention; In the research, it is highlighted it is possible to contribute by increasing the scope of studies that work the family perspective in front of the child with biliary atresia; and in care, one can contribute through reduction of anxiety and stress from a list of care can be oriented to these family members.

Method

This is an integrative literature review, according to Mendes et al., ^(7,8) who point out six steps for the elaboration of an integrative review, these are: (I) Definition of the review question; (II) search and selection of primary studies; (III) data extraction from primary studies; (IV) critical evaluation of primary studies; (V) synthesis of the results of the review and (VI) presentation of the review.

For the guiding question elaboration, PVO (Population, Variables and Outcomes) strategy was used, being thus, discriminated: population – family members; variables – biliary atresia; outcome – health care. This strategy allowed the elaboration of the following guiding question: “What is the knowledge produced about the impact of biliary atresia on

the child and the family?”. The Table 1 summarizes how descriptors were identified from PVO strategy.

Table 1. PVO strategy. DeCS descriptors for the components of the guiding question

Strategy items	Components	Descriptors
Population	Family	Cuidadores/caregivers Família/family
Variables	Biliary atresia	Atresia biliar/biliary atresia
Outcomes	Care	Cuidado da criança/child care Criança/child or children

Source: Own elaboration (2022)

The search for the articles was conducted between July and September 2022. This stage is responsible for establishing search strategies in databases and the studies' inclusion and exclusion criteria. Databases and libraries searched were SCOPUS, PUBMED and EBSCO. These databases were chosen due to their scope of scientific relevance. Search strategies used were made from the descriptors in English and Portuguese combined with the Boolean operator AND, according to the Table 2.

Table 2. Systematization of searches in databases/data libraries

Base/biblioteca	Search 1	Search 2
SCOPUS	caregivers AND biliary atresia	
PUBMED	child care AND biliary atresia and family	caregivers AND biliary atresia
EBSCO	child AND biliary atresia AND family	AND child care subject AND biliary atresia allfields

Source: Own elaboration (2022)

The following filters were used: full text, published in the last five years. As inclusion criteria in all databases, studies presenting results on the theme were included and literature reviews, abstracts, reviews, monographs, dissertations, repeated studies and duplicates were excluded.

To select the studies, we used Microsoft Excel, which were selected in a careful and systematized way.

For data extraction, we used a form prepared by the author based on the instrument of Mendes et al.,^(7,8) containing the following variables to be collected: origin/journal and volume, title, author and year, objective, main results and methodological design, which are presented in a synoptic table.

Studies included in the sample were classified according to their level of evidence. According to Melnyk and Fineout-Overholt⁽⁹⁾ the evidence can be classified into: (I) Level

1: Controlled or randomized clinical trials, which used meta-analysis; (II) Level 2: Evidence from studies with experimental design; (III) Level 3: Evidence from a study with a quasi-experimental design; (IV) Level 4: Evidence from non-experimental, descriptive or qualitative studies; (V) Level 5: Evidence resulting from case reports or experience; and (VI) Level 6: Evidence from expert opinions.

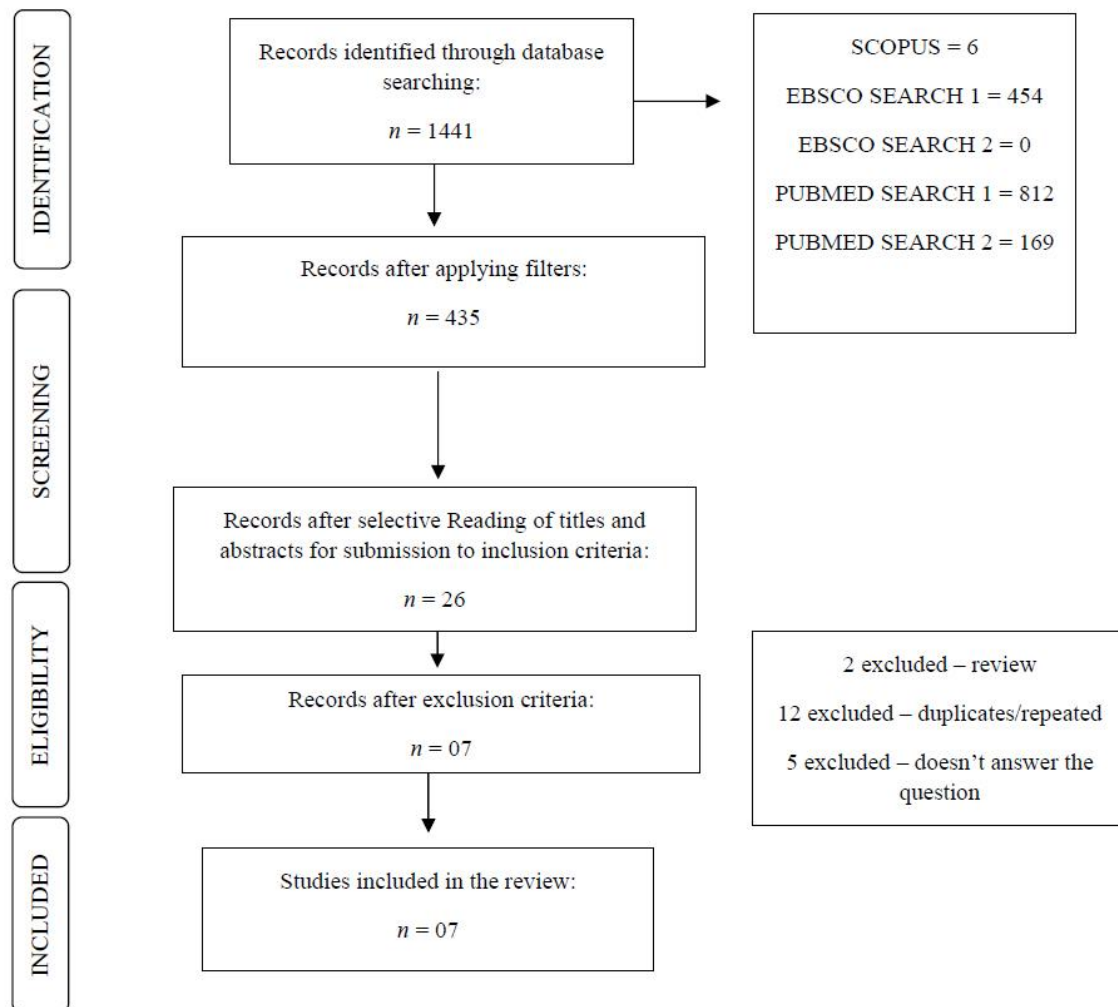
Results were synthesized, discussing them with pertinent literature, identifying the gaps in studies' knowledge and the review' limitations, as well as making recommendations for practice.

Regarding the ethical procedures of this research, it did not need to be forwarded to the Ethics and Research Committee, since it is a review of studies already publicly available. The authors of this study also report that they followed copyright law no. 9,610 of February 19, 1989. ⁽¹⁰⁾

Results

In the general search, a total of 1441 articles were found, which after applying the full-text filters freely available and published in the last five years, obtained the quantity of 435 articles, which went through the reading of title and abstract to apply the inclusion criteria. After reading the titles and abstracts, 26 studies were included for full reading, of which 7 are part of this review and were analyzed qualitatively.

Figure 1. PRISMA with schematization of the articles' search



Source: Adaptation of PRISMA.⁽¹¹⁾

Table 3 presents the synopsis of the studies found as well as the main results and level of evidence of the studies. Regarding the main results, there were divided into: impact for the child and impact for the family, in which not all studies presented results pertinent to both aspects.

Table 3. Synopsis of the included studies

Origin Journal Volume	Title	Author and year	Objective	Method	Main results	Level of evid.
EBSCO Journal of Pediatric Gastroenterology and Nutrition v. 71	Quality of life in parents of children with biliary atresia	Rodijk et al. 2020	To determine quality of life (QoL), stress, and anxiety levels in parents of children with biliary atresia (BA), and to assess factors associated with parental quality of life.	Cross-sectional study with a qualitative approach conducted with 61 parents of children with biliary atresia	Impact for the child	IV
					Parents rate the health of their children with biliary atresia as regular to poor.	
					Impact for the family	
	Emotional impact and reduction of time to perform family activities, because the time was destined to the child care.					
PUBMED World Journal of Clinical Cases v. 09	Application of narrative nursing in families of children with biliary atresia: retrospective study	Zhang et al. 2021	To explore the application value of narrative nursing in the families of children with BA.	Quantitative, experimental study conducted with 64 families of children with biliary atresia. 32 families received routine nursing (control group) and 32 families received narrative nursing care (study group).	Impact for the family	IV
					The parents had anxiety and depression related to drastic life change to care for their child with biliary atresia.	
PUBMED Journal of Clinical Medicine v. 11	Variability of care and access to transplantation for children with biliary atresia who need a liver replacement	De Goyet et al. 2022	To compare the patients who were initially seen with those who were immediately referred for diagnosis and seen at the center that eventually offered analysis of intention to transplant.	Multicenter retrospective analysis with a quantitative approach, performed with 219 children with biliary atresia.	Impact for the child	IV
	Complications caused by biliary atresia reflect a poor perception about the health of children with this disease.					
PUBMED BMC Pediatrics v. 20	Feeding practices in 6–24 month-old children with chronic cholestatic liver diseases: a mixed-method study	Chen et al. 2020	To evaluate the feeding status of chronic cholestatic children aged 6–24 months and to explore their parents' experiences with feeding practices.	Mixed study conducted with 74 children.	Impact for the family	IV
	Concerns about the most appropriate diet for the child's condition.					

PUBMED Orphanet Journal of Rare Diseases v. 16	Quality of life and mental health of children with rare congenital surgical diseases and their parents during the COVID-19 pandemic	Fuerboeter et al. 2021	To evaluate the health-related quality of life (HRQoL), quality of life (QoL), and mental health of children with rare congenital surgical diseases and their parents during the COVID-19 pandemic and lockdown measures.	Cross-sectional observational study with a quantitative approach, conducted with 298 parents.	Impact for the family	IV
					Impairment of mental health, especially among mothers, when compared to the mental health of fathers.	
PUBMED Orphanet Journal of Rare Diseases v. 16	Perceived mental health in parents of children with rare congenital surgical diseases: a double ABCX model considering gender	Boettcher et al. 2021	To investigate factors within the Double ABCX model to explain parental mental health when raising a child with a rare congenital surgical disease.	Cross-sectional study with a quantitative approach, conducted with 210 parents.	Impact for the family	IV
					Impaired family functioning and social support and increased stressors.	
SCOPUS Translational Pediatrics v. 10	Factors influencing quality of life in children after biliary atresia treatment	Liang et al. 2021	To investigate the current quality of life of children after biliary atresia treatment.	Cross-sectional study with a quantitative approach was conducted with 70 children with biliary atresia.	Impact for the child	IV
					Children with biliary atresia have reduced physical and cognitive health when compared to children without biliary atresia.	
					Impact for the family	
	Caregivers of children with biliary atresia presented fatigue and depression.					

Source: Own elaboration (2022)

Methodological aspects of the studies

Most studies presented a quantitative approach ($n=5$), followed by two studies with a qualitative and mixed approach, respectively. With regard to quantitative research, it is characterized by the evaluation of a theory, associated with quantified variables and data, which are presented through numbers and statistics, defining whether the assumptions presumed in the theory are supported or not. ⁽¹²⁾

About qualitative research, Melnyk⁽⁹⁾ approaches it makes it possible to know the nature of the meanings, the motives, the beliefs, the values, the attitudes, among other characteristics of a given problem in question. Still, it is concerned with a level of reality that cannot be measured or quantified, but enables a process of change of social reality.⁽⁹⁾ And about a mixed method, it compiles the collection and integration of quantitative and qualitative data in the same research, so one complements the other.^(13, 14)

Discussions

Impacts of biliary atresia for the child

The study by Rodijk et al.⁽¹⁵⁾ conducted with 61 parents of 39 children with biliary atresia (31 of these children underwent liver transplantation), identified 16 % of parents evaluated the health of their children as regular and 5 % evaluated as poor. It is important to integrate the result of the previous study with findings of the study by De Goyet et al.,⁽¹⁶⁾ since complications caused by biliary atresia reflect significantly on perception of health. Thus, this study was conducted with 219 children with biliary atresia, of which 39 required liver transplantation, in which it was found the fact they were older children caused complications such as cirrhosis to have already been developed.

The study by Ferreira et al.⁽¹⁷⁾ reveals absence of bile flow and age of Kasai surgery greater than 90 days can predict death or need for liver transplantation. It should be noted due to rapid evolution to liver failure, biliary atresia is the major cause of liver transplantation in children,⁽¹⁸⁾ and it is therefore necessary to follow up this child even after the Kasai procedure.

The study by Liang et al.⁽¹⁹⁾ presents biliary atresia brings negative repercussions to quality of life of affected children, in which after surgery for treatment of this disease, the children of the study presented significantly lower levels of physical health, emotional function and cognitive function when compared to healthy children of the same age, which corroborates with the study of Araújo et al.⁽²⁰⁾ It is important to highlight health conditions that require invasive procedures as treatment is usually exhausting for the child and family members, reflecting significantly on the mental state of those involved.⁽²¹⁾ In addition, children's quality of life after surgery was related to coexisting diseases, knowledge about liver transplantation, form of payment of medical expenses and duration of antibiotic therapy.⁽¹⁹⁾

Impacts of biliary atresia for the family of the affected child

It was evidenced that parents of children with biliary atresia scored higher in the psychological domain of quality of life compared to reference data from the general population, had emotional impact and time reduction, impacting on the reduction of family activities. However, data on family cohesion were more positive in relation to the parents of children with biliary atresia when compared to the general population.⁽¹⁵⁾

In the interventional research of Zhang et al.⁽²²⁾ verified the effect of narrative nursing on the parents of children with biliary atresia, they applied scales of self-assessment of anxiety and depression, identifying before intervention both control and intervention group presented symptoms of anxiety and depression, in addition to having to adjust the following aspects: modify their life to match the child's needs, assess family and social resources meet the child's needs, deal with personal emotions, and adapt to the role of caregiver to care for the child with biliary atresia.

In the study by Chen et al.,⁽²³⁾ conducted with 74 children with chronic liver diseases (16 of whom had biliary atresia), identified a low percentage (16 % of children) of adequate nutrition. The vast majority of children had inadequate nutrition, and family members reported lack of knowledge about feeding (quantity, variety and age of food introduction), misunderstanding about feeding process (they consider children should have a differentiated diet, as smooth as possible being influenced by the opinion of family and friends) and lack of knowledge about the importance of complementary foods (parents believed milk has higher nutritional value and did not give importance to complementary feeding).

Usually it is still quite common for parents to present ignorance about healthy eating for the child,⁽⁶⁾ and when it comes to children with biliary atresia this problem increases, since it is common due to the atresia of the bile ducts there is greater gastrointestinal loss, impairing nutrients absorption and requiring caregivers to pay attention to signs of malnutrition and act on vitamin and nutrient supplementation.⁽²⁴⁾

In the research of Fuerboeter et al.,⁽²⁵⁾ conducted with 210 parents of children with rare congenital surgical diseases (index group with 14 children with biliary atresia) and with 88 parents of children without previous diagnoses or signs and symptoms of rare congenital surgical diseases (control group with 20 children with atresia), it presents the quality of life and mental health of the parents. In the index group, parents had much lower quality of life than the control group. Regarding mental health, mothers generally showed greater impairment in all mental health scores compared to fathers. Being male was associated with lower health impairment, while women with lower schooling and who had a high level of care were associated with low quality of life and greater impairment to mental health, respectively.

Greater impairment of the mothers' mental health may be related to the fact they are the most present caregivers. This is due to the culture of sexual division of labor, in which it is up to the woman to take care of children, often taking responsibility and concerns only for herself. In addition, the woman must reconcile domestic care with the child's care that demands special attention due to its health condition, which can be related to the overload and impact on the mental health of the mother.^(18, 26)

In addition, it is highlighted the main agent behind the decisions about medical treatment are the parents and it is also up to them to work for the gradual transfer of care when puberty arrives, also aiming to strengthen autonomy and not harm the follow-up of treatment if it extends for so long.^(16, 23)

In addition, families need to assist or supervise the child to dress, perform hygiene and also provide food and leisure, since development of children with biliary atresia is impaired.⁽¹⁸⁾ Still, taking into account chronic diseases usually impact on the child's development, this is another concern family members need to deal with.⁽²⁰⁾

In this context, until the children can take care of themselves, parents begin to adjust their lives to meet the child's care needs,⁽²²⁾ failing to take care of their own health to the detriment of the well-being of the offspring.

Given so many concerns, it is important parents and/or caregivers are trained and oriented to care for the child. The study by Lima-Alves et al.⁽²⁷⁾ highlights the importance of preparation of the parents throughout child hospitalization, with regard to their home care, especially with regard to antibiotics' administration, with guidance on medical prescription, times of drug's administration, using a more understandable language, in addition to demonstrations.

In another research conducted with 210 parents of children with rare congenital surgical diseases (14 of whom were parents of children with biliary atresia), it was evidenced

family functioning, social support and perceived stressors are directly associated with the mental state despite the parents' coping with the child's disease.⁽²⁸⁾ Study by Liang et al.⁽¹⁹⁾ presents caregivers of their study manifested fatigue and depression and felt unbalanced, lonely and revealed they were easy to get angry. This occurs due to the disease's unpredictability and as a result of the parents feeling powerless.⁽¹⁸⁾

Families of children with chronic illnesses are disconnected from their daily lives and often have no control over their own lives while living with chronic illnesses. Notably, when families are fully informed and trained throughout the life of the chronically ill, receiving care from health and education professionals, it is easier to adjust to the new situation of the child, minimizing the negative impacts.⁽¹⁸⁾

In this context, information provided by the nursing team to the parents becomes essential to promote and facilitate their participation in child's care. And this partnership in child care between nursing-family shows benefits for both the child and its family, because the opportunity to participate in care, reduce anxiety levels, increase the feeling of independence and promote adequate communication, stimulating and facilitates adaptation to the new health condition of the child.⁽²¹⁾ Thus, narrative nursing gains prominence, while it manages to inform family members and integrate them into the child's care.

It is worth mentioning a nurse contributes to the surgical care of the child through the Systematization of Nursing Care which enhances surgical care, but its role is not limited to this.⁽²⁹⁾ The nurse also, based on family-centered care, can use light or relational technologies both in hospitalization scenario and in the context of primary care, aiming to create a bond with the parents/caregivers, with a view to guiding them about the care the child will require at home and how caregivers can optimize child's quality of life, minimizing risk of infections and promoting the necessary care for child's full development.⁽⁶⁾ Also, for children over 3 years old, it is already possible to implement the therapeutic toy by the nurse. And in addition, this professional can teach parents how to implement play at the time of child care.^(20, 30)

A limitation of this study is the fact most studies focus on pathological, pharmacological, therapeutic and diagnostic aspects of the disease, making it difficult to analyze aspects related to the family and the child. Future studies should approach the child with biliary atresia from an integral perspective, considering family members' difficulties and the impacts of the disease and treatments on the quality of life of the child and their caregivers.

Final considerations

Impacts of biliary atresia for affected children observed in the studies relate to disease's consequences regarding need for surgical procedure (Kasai surgery or transplantation) negatively impact child's quality of life. Impacts on family are related to consequences for mental health and low quality of life combined with lack of knowledge about child care and excessive worry.

Nurses can contribute significantly from their informational competence, by using educational technologies to guide on child's feeding, administration of antibiotics, as well as on management of emotional crisis of the family that takes care and worries for this child, which demonstrates the relevance of this research.

The present study has potential to stimulate new research and contribute to nursing being responsible for promoting health of children and family members, contributing to health education actions to provide guidance on necessary care, which reduces anxiety and fear.

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