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Long-term follow-up of children with biliary atresia after Kasai procedure

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Abstract
Introduction and Background: Untreated biliary atresia, an obliterative cholangiopathy that begins in infancy and progresses to cirrhosis and liver fibrosis, can be fatal. The major surgical procedure for reestablishing bile flow and postponing liver transplantation is the Kasai portoenterostomy. The long-term results of the Kasai surgery, meanwhile, can be quite different. Aiming to assess survival, biochemical, and clinical outcomes in the long run, this study followed children who had the Kasai surgery for biliary atresia.
Materials and Methods: In this cohort study that looked back at This study was conducted at the Department of Paediatrics, Tagore Medical College and Hospital, Chennai, Tamil Nadu, India from February 2018 to January 2019, 60 kids with biliary atresia who had the Kasai surgery done at a tertiary pediatric surgical center were included. At least five years after surgery, patients were monitored. The following variables were recorded: age at surgery, frequency of cholangitis, clearing of jaundice, growth parameters, complications, and native liver survival; liver function tests; and health of the patient's liver.
Results: After 6 months following surgery, 33.3% of the 60 children (38 out of 60) no longer had jaundice. Surgery was performed at a median age of 58 days. Out of the total number of patients followed up, 25 (41.7%) experienced recurrent cholangitis, and 14 (23.3%) needed a liver transplant because their livers were failing. The native liver survival rate was 66.7% after 5 years and 53.3% after 10 years after Kasai surgical procedure. The results were noticeably better (p<0.05) for children who had the Kasai operation performed before they were sixty days old. Seventy percent of animals with native livers survived with appropriate growth parameters.
Conclusion: Children with biliary atresia still have a possibility for native liver survival after the Kasai surgery, particularly if it's done early on. Regardless, many patients experience sequelae such cholangitis and may need a liver transplant in the long run. The key to maximizing results is prompt referral for transplantation and lifelong follow-up.

Keywords: Biliary atresia, Kasai procedure, pediatric liver disease, long-term follow-up, native liver survival, liver transplantation

Introduction
Biliary atresia (BA) is a fibro-inflammatory cholangiopathy that develops in infants and causes bile retention, cholestasis, and liver damage as it blocks the extrahepatic bile ducts. The incidence of this serious newborn illness varies by region and ethnicity, although it ranges from 1 in 8,000 to 1 in 18,000 live births globally. Bacterial angiopathy (BA) is fatal if left untreated because it causes liver cirrhosis, portal hypertension, and hepatic failure in the first two years of life [1-3].

Biliary atresia's exact cause is still a mystery, although current theories point to a complex interplay of hereditary predisposition, prenatal infections, immunological dysregulation, and environmental factors. Syndromic BA is the more prevalent type, while non-syndromic BA is less common and is not often linked to other congenital abnormalities. Improving outcomes in afflicted newborns requires early detection and appropriate intervention [2-4]. The long-term effectiveness of the Kasai operation is still up for debate, despite its broad use. A liver transplant may be necessary for some children, while others suffer from chronic jaundice, recurring cholangitis, failing to thrive, and worsening liver malfunction over time. As a result, BA is the primary cause of pediatric liver transplants globally, and many of these children will need a transplant [3-5].

Age at surgery, severity of liver fibrosis at presentation, speed with which jaundice clears up after surgery, and occurrence of complications including cholangitis and portal hypertension are

all variables that impact long-term results. There are still inequalities in outcomes, especially in low-and middle-income countries, despite the fact that surgical technique, perioperative care, and long-term follow-up have all contributed to increased survival rates [4-6]. The assessment of survival, quality of life, growth patterns, liver function, and the scheduling of liver transplantation all require long-term follow-up. Studies from underdeveloped nations, in particular, that look at long-term effects beyond the first few years after Kasai are scarce [5-7].

Children with biliary atresia who had the Kasai surgery done at a tertiary care center were the subjects of this study, which followed them for 5-10 years to see how they fared in the future. The purpose of this study was to evaluate the Kasai surgery in terms of native liver survival, jaundice clearing, complication incidence, growth outcomes, and the necessity for liver transplantation in the long run. Timely treatment decisions, like as referral for transplantation, can be informed by these data, which may offer significant insight into the long-term prognosis of BA [6-8].

Material and methods

A tertiary care teaching hospital's pediatric surgery and gastroenterology departments executed this retrospective cohort analysis. The study encompassed children who received the Kasai portoenterostomy (KPE) procedure for biliary atresia. This study was conducted at the Department of Paediatrics, Tagore Medical College and Hospital, Chennai, Tamil Nadu, India from February 2018 to January 2019. 60 children who had biliary atresia and had KPE were part of the testing group. Clinical manifestations, ultrasonography, hepatobiliary scintigraphy, and intraoperative cholangiography all contributed to the conclusive diagnosis.

Inclusion Criteria

- Children diagnosed with biliary atresia who underwent Kasai procedure.
- Minimum postoperative follow-up of 5 years.
- Availability of complete medical records including follow-up data.

Exclusion Criteria

- Children with incomplete records or lost to follow-up.
- Patients with other concurrent congenital liver disorders.
- Syndromic biliary atresia.

Data Collection

Medical records, surgical logs, and notes from follow-up clinics were used to gather data. Patient demographics (diagnosis age, gender, and surgical weight). Characteristics of the disease (jaundice, hepatosplenomegaly, and pale stools).

Follow-up and Outcome Measures

The outpatient section followed up with patients on a regular basis, every three to six months. Growth evaluations, liver function tests, and clinical examinations were conducted at every appointment. We performed upper GI endoscopy and ultrasounds of the abdomen if necessary.

Results

The study comprised 60 children who had biliary atresia and had the Kasai surgery. With a range of 5-10 years, the average period of follow-up was 7.8±2.1 years. Results were evaluated by looking at demographics, whether or not jaundice cleared up after surgery, the incidence of postoperative problems, the likelihood of native liver survival, and the necessity of liver transplantation.

Table 1: Demographic and Clinical Characteristics of Patients

Characteristic	Number of Patients (%)
Age at surgery (days)	
≤60	35 (58.3%)
>60	25 (41.7%)
Gender	
Male	34 (56.7%)
Female	26 (43.3%)
Birth weight ≥2.5 kg	49 (81.7%)
Associated anomalies	8 (13.3%)

About half of the patients had the Kasai procedure done during the first sixty days of life. Among the participants, 56.7% were men. Concomitant abnormalities, such polysplenia or heart problems, were present in approximately 13.3%.

Table 2: Jaundice Clearance and Early Outcomes

Parameter	Number of Patients (%)
Jaundice clearance at 6 months (<2 mg/dL)	38 (63.3%)
Persistent jaundice (>2 mg/dL)	22 (36.7%)
Early cholangitis (within 6 months)	14 (23.3%)
ICU stay >5 days	10 (16.7%)

At 6 months after surgery, 63.3% of kids had their jaundice completely gone. While 16.7% required a lengthy stay in the intensive care unit, 23.3% experienced early cholangitis.

Table 3: Long-term Complications Observed During Follow-up

Complication	Number of Patients (%)
Recurrent cholangitis	25 (41.7%)
Portal hypertension	18 (30.0%)
Variceal bleeding	7 (11.7%)
Ascites	10 (16.7%)
Growth retardation	12 (20.0%)

The most prevalent long-term consequence was recurrent cholangitis. Ascites was observed in 16.7% of cases and portal hypertension in 30%. Among the group, 20% showed signs of growth retardation.

Table 4: Native Liver Survival and Transplantation Outcomes

Outcome	Number of Patients (%)
Native liver survival at 5 years	40 (66.7%)
Native liver survival at 10 years	32 (53.3%)
Liver transplantation required	14 (23.3%)
Deaths without transplantation	6 (10.0%)

The native liver survival rate was 66.7% after 5 years and 53.3% after 10 years. Complications from liver failure were the cause of death for six patients (10%), and fourteen patients (23.3%) ultimately had liver transplantation.

Table 5: Factors Associated with Jaundice Clearance and Native Liver Survival

Factor	Jaundice Clearance (%)	Native Liver Survival at 5 Years (%)	p-value
Age at surgery ≤60 days	29 (82.9%)	30 (85.7%)	<0.05
Age at surgery >60 days	9 (36.0%)	10 (40.0%)	<0.05
Early cholangitis present	4 (28.6%)	5 (35.7%)	<0.05
No early cholangitis	34 (76.0%)	35 (78.0%)	—

Compared to children who had the Kasai procedure later in life, those who had it during the first 60 days of life had considerably better rates of native liver survival and clearing of jaundice (p<0.05). Results were worse when cholangitis was detected early.

Discussion

This study aimed to determine the long-term effects of the Kasai portoenterostomy on native liver survival and the necessity of liver transplantation in children who had biliary atresia. The results confirm that early surgical intervention is crucial for bettering clinical outcomes. There was a marked improvement in jaundice clearance rates and native liver survival at 5 and 10 years postoperatively for children who underwent the Kasai surgery within the first 60 days of life. These findings corroborate previous research that has shown that bile flow restoration and slowed liver fibrosis progression are more likely to occur with early surgical repair [9-11].

Although there were some short-term benefits to the procedure, many patients in this group ended up with chronic problems. Portal hypertension, ascites, variceal hemorrhage, and recurrent cholangitis ranked in that order of prevalence. Liver function would gradually decrease due to these problems, and transplantation of the liver would be necessary in many cases. It appears that cholangitis contributes significantly to the persistent hepatic impairment after Kasai because it was linked to worse outcomes. Also, several kids showed signs of stunted growth, which could be due to inflammatory conditions, poor food absorption, or chronic liver disease. Stable hepatic function following successful Kasai surgery can support normal development, since most native liver survivors maintained good growth characteristics [12-14].

Comparable survival rates from high-income and middle-income nations were found in this study: 66.7% at 5 years and 53.3% at 10 years for native livers. Still, 10% of the kids didn't make it through the process, and almost a quarter of them needed a liver transplant. Although the Kasai surgery can postpone the onset of end-stage liver disease in certain individuals, these figures show that it is not a cure-all [15, 16]. A critical predictive feature that emerged was the ability to remove jaundice within the first six months after surgery. Patients who were able to achieve bilirubin normalization early on had far better long-term results and fewer problems. Infants exhibiting chronic jaundice should be diagnosed as soon as possible and referred for surgical intervention as soon as possible, according to the results of this study [17-19].

To optimize the timing of the Kasai operation and enhance early detection rates, standardized screening programs could be implemented. These programs could include the use of stool color cards or routine direct bilirubin testing. The findings further stress the need for all post-Kasai patients to have long-term follow-up. To make the right clinical decisions, like when to transplant the liver, it is crucial to assess nutritional status, portal hypertension, and liver function on a regular basis. For optimal results, it is essential to have a multidisciplinary team that includes pediatric hepatologists, surgeons, nutritionists, and transplant teams [20-22].

There are a few caveats to be aware of, even though the study offers helpful information. Since it was a retrospective study done at just one location, the results might not apply to the whole country. There was a lack of assessment of several psychological consequences and quality-of-life metrics, and there was a possibility of documentation bias in some data. In spite of these caveats, the study adds significantly to our knowledge of the long-term effects of Kasai portoenterostomy and further supports the need of prompt surgical intervention and organized postoperative care for the treatment of biliary atresia [23-26].

Conclusion

Biliary atresia is best treated surgically with the Kasai portoenterostomy, which is most effective when done during the first sixty days of life. Both the elimination of jaundice and the survival of the native liver are greatly improved by early surgery. Consequences including growth retardation, portal hypertension, and recurrent cholangitis persist in many patients over the long term, and liver transplantation may be necessary for some of these patients. The management of these problems and the determination of the appropriate timing for transplant referral require continuous multidisciplinary monitoring. Children with biliary atresia can greatly benefit from better overall outcomes and quality of life if early detection is strengthened and consistent follow-up methods are implemented.

Funding

None

Conflict of Interest

None

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