Pattern and Survival of Biliary Atresia Patients; Experience in Southern Nigeria

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ABSTRACT

Background: Biliary atresia (BA) has been a challenge to surgeons worldwide. Beyond the revolutionary surgical technique popularised by Kasai, liver transplantation has added renewed hope in the long-term outcome. In Nigeria, where late presentation is very common, there is need to assess the long-term results of the treatment options available to us. **Aim:** We aimed to evaluate the presentation and management outcome of BA and the long-term survival of BA patients seen in our practice. **Materials and Methods:** Cases of BA seen between January 2007 and December 2011 in three tertiary health facilities in South East Nigeria were included. Data obtained included age at presentation, clinical features, treatment offered and age at the time of death. Analysis was with the SPSS 17.0. **Results:** Twenty four patients comprising 10 (41.7%) males and 14 (58.3%) females were included in the study. The mean age of presentation was 4.02 (±214) months; range 1.75-11.0 months. Fifteen (62.5%) patients had surgery while 9 (37.5%) received medical treatment only. The mean age at death was 14.2 (±8.1) months; range 2.5-30 months. **Conclusion:** BA poses a daunting challenge in our practice. Outcome of treatment is still discouraging. We identified late presentation, lack of facilities to make early diagnosis, lack of adequately trained manpower to manage these children and lack of post-operative care and support for patients as the major challenges in the management of BA children in our region.

KEYWORDS: Atresia, biliary, pattern, surgical correction, survival

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Access this article online	
Quick Response Code:	Website: www.nigerianjsurg.com
	DOI: 10.4103/1117-6806.111495

INTRODUCTION

Biliary Atresia (BA) is among the most common clinical conditions leading to cholestasis and liver transplantation in children.[1,2] It is uniformly fatal, if untreated. The treatment of BA was revolutionized by the introduction of liver transplantation in addition to porto-enterostomy originally described by Kasai.[3] In recent times, the outlook of BA treatment has been in the upward trend in many advanced countries.[4] Some centers have reported survival of patient to adulthood without the need for liver transplantation.[5,6] The European BA registry reported an overall survival of 78%.[7] Conversely, reports of poor outcome of treatment are rife in many developing countries where late presentation is a continuing problem. In addition, distinguishing BA from other causes of neonatal jaundice and cholestasis in the early phases in such resource poor environments has been quite a challenge for clinicians. The outlook of BA in the West African sub-region appears rather bleak.[8,9] Currently, there are limited published reports on the presentation, management outcome and life expectancy of patients of BA in Nigeria. This study is to assess the presentation of BA and long-term survival of patients of BA seen in three tertiary health facilities in South Eastern Nigeria. It is hoped to highlight the challenges in the management of BA in South Eastern Nigeria and suggest ways of improving on them.

MATERIALS AND METHODS

This study was carried out with the approval of the ethical committees of our institutions. Informed consent was obtained from the parents of the children who were included prospectively. Cases of BA seen by the authors between January 2007 and December 2011 were included in this study. The inclusion criteria were: History of progressive jaundice in an infant, ±pale (clay) stools, conjugated hyperbilirubinemia, absent or small gall bladder and extrahepatic ducts ± hepatomegaly. Exclusion criteria were loss to follow-up and incomplete data. Data was obtained both retrospectively and prospectively. Phone contacts of parents and guardians were obtained to enhance follow-up. Data obtained were: Age, gender, presenting clinical features, any previous treatments, ultrasound and liver function test reports, findings at surgery, medical and surgical treatment

offered, response to treatment, survival outcome. Data was collated and analyzed with the 17.0 version of the Statistical Package for the Social Sciences (SPSS® Chicago, IL). Test of significance was done with the Student *t*-test. Significant level was taken at *P* value of 0.05.

Results

A total of 31 consecutive cases of BA were seen over the 5-year period of the study. Seven (22.6%) patients were excluded due to incomplete data or loss to follow-up. Twenty four (77.4%) patients were followed-up until they died. There were 10 (41.7%) males and 14 (58.3%) females (M:F = 1:1.4). Six (25%) patients presented within 3 months of life but the rest presented after 3 months [Table 1]. The mean age at presentation was 4.02 (±2.14; range 1.7-11.0) months. All patients had jaundice, hepatomegaly, deranged liver function, absent or hypoplastic gall bladder, and umbilical hernia at presentation. Twenty two (91.7%) passed clay colored stool and 4 (16.7%) had inguinal hernia. Biopsy of the liver supported diagnosis in all cases. All patients received a combination of steroids, antibiotics and cholestyramine. Surgery was not possible in 9 (37.5%) patients because of financial constraints, refusal of consent by parents, or lack of fitness for surgery. Of the patients who were operated on (n = 15), the liver was enlarged, hard and craggy in 9 (60%), [Figure 1], Kasai operation was performed in 6 (40%), choledochojejunostomy in 2 (13.3%), exploratory laparotomy without any shunt in 7 (46.7%). Of the operated patients, 5 (33.3%) who had Kasai operation improved with reduction in jaundice for a period of 2-4 months, 8 (53.3%) did not improve, and 2 (13.3%) died from surgery related complications. A total of 19 (79.2%) patients survived to the age of 6 months but none attained the age of 36 months. The immediate cause of death was portal hypertension and bleeding in 8 (33.3%), hepatic failure in 3 (12.5%). Cause was not established in 11 (45.8%) patients who died outside the hospital. The overall mean age of death of all the patients was 14.2 (±8.1; range 2.5-30) months. Table 2 shows the duration of life of the patients followed-up. There was no statistical difference between the survival of patients who were operated on and those who were not (P = 0.27).

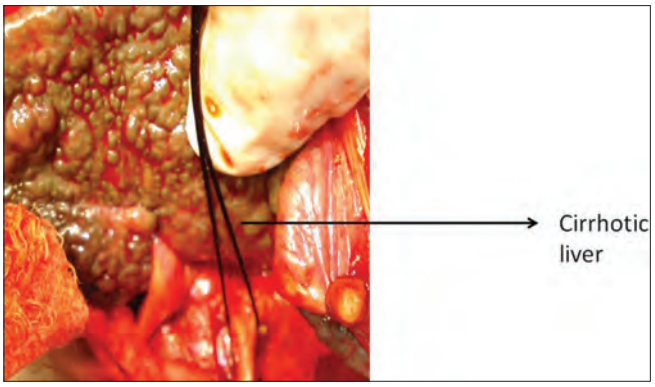

Figure 1: Absent gall bladder; cirrhotic liver

Table 1: The age of patients at presentation

Age at presentation (months)	Patients (n=24) (%)
<3	6 (25)
3-6	11 (45.8)
6-9	4 (16.7)
>9	3 (12.5)

Table 2: Ages attained by biliary atresia patients who received medical treatment and surgery versus patients who received only medical treatment

Age attained before death (months)	Patients according to treatment	
	Medical+surgical (n=15) (%)	Medical only (n=9) (%)
6	11 (73.3)	8 (88.9)
12	8 (53.3)	6 (66.7)
18	4 (26.7)	3 (33.3)
24	3 (20.0)	2 (22.2)
30	1 (6.7)	0 (0)
36	0 (0)	0 (0)

P=0.78

Discussion

The results in this study showed that the majority of our patients were presented after 3 months of life. At this time, severe liver derangement had already occurred in most of them. Though, most of the delays occur at the primary health-care level, at the tertiary level, diagnosis has sometimes been elusive or delayed due to inadequate facilities to investigate and confirm diagnosis. Our treatment protocol for all BA patients has been surgery, where possible, and a triple therapy of antibiotics, low-dose steroid and cholestyramine. The use of steroids in the management of these patients is common though its effectiveness is still debated.[10,11] Currently we offer surgery to all patients presenting to us with BA irrespective of the age of patient. This is because we see many of our patients presenting after 3 months of age and we consider surgery their only chance of survival. More so, some authors have reported good results even in patients older than 3 months of age.[1,6,12] However, this practice results in a high-rate of open and close surgery where patients have been explored but bile shunting was not possible due to badly cirrhotic liver. Our patients who had shown improvement following surgery, deteriorated again within 12 months of surgery. These patients ostensibly, developed ascending cholangitis with subsequent portal fibrosis or liver cirrhosis. At this stage, liver transplantation would have been the option for the patients but this service is not yet available in Nigeria. Efficient follow-up and early detection of complications in patients who have had surgical correction of BA or liver transplantation is known to enhance their survival.[13] The lack of such efficient follow-up and support may be contributory to the shortness of long-term survival of our patients. The absence of any statistical difference between the survival of our patients who were subjected to surgery and those who were treated medically is difficult to interpret since the patients were not matched for age at presentation and type of BA. This study has however shown that the survival of our BA patients is less than

3 years with or without surgical correction. This poses a serious ethical question as to whether there is sufficient justification in attempting surgical correction in these patients in our practice. We believe that this outlook of BA in Nigeria is similar to what is obtainable in many other developing countries. Sanghai *et al.*,[14] have also reported similar experience of late presentation and 33% incidence of improvement with surgical correction in their series from Western India.[5,14] Encouraging reports of successful care of these patients come from centers where there are concerted efforts by all concerned to improve quality of care. Many researchers have contributed further to these successes by identifying prognostic factors affecting outcome in BA patients and employing such knowledge to improve their results.[15] Liver transplantation for patients who already have cirrhosis is now shifting from the conventional technique to micro-surgical biliary reconstruction in some centers with attendant better results.[16] We identify late presentation, lack of facilities to make early diagnosis, lack of well-trained manpower to manage these children, and lack of post-operative care and support for patients, as the major challenges in the management of BA children in our region. There is need to put measures in place to make for earlier diagnosis of BA. A protocol of liver scanning, and serial liver function test for all babies with jaundice at all levels of care is needed. Workshops and seminars both at primary, secondary and tertiary healthcare levels will raise the index of suspicion among healthcare practitioners. Our hospitals records indicate that we see about 2-6 cases of BA per annum. This load of patients is too low and does not make for improved skill and experience of the teams managing these patients. There may be need to establish regional biliary surgery centers where all babies suspected of BA are referred and managed by teams specially dedicated to management of BA. This concept of regional biliary centers has been put into practice in some advanced countries and it has been shown to improve the management outcome and life expectancy of these children.[4,17]

Conclusion

BA is a major challenge in our practice in Nigeria. Lateness of presentation and diagnosis are major contributory factors. Most patients have already developed liver cirrhosis by the time diagnosis is made. Interventions presently available in our practice are not sufficient to prolong life or significantly improve the outlook. The outcome of treatment in our experience therefore raises ethical questions on the usefulness of surgical treatment in our patients. There is need for a coordinated plan of action among health-care givers in our region to achieve improved results. Centers dedicated to the management of these patients will make for a better articulation of service delivery. Larger loads of cases in such dedicated centers will also enhance the experience of the surgeons and ultimate results of treatment.

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How to cite this article: Okoro PE, Igwe P, Opara PI. Pattern and survival of biliary atresia patients; Experience in Southern Nigeria. Niger J Surg 2013;19:4-6.

Source of Support: Nil. **Conflict of Interest:** None declared.