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Biliary Atresia – An Easily Missed Cause of Jaundice amongst Children in Uganda

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Back ground: ***Biliary atresia is characterized by biliary obstruction, it has an incidence of 1:15000 and presents with jaundice, acholic stools / dark urine and hepatomegaly. This disease rapidly leads to liver cirrhosis and liver failure if untreated surgically. The main objective was to establish the epidemiology of patients presenting with biliary atresia and immediate surgical outcome.***
Methods: ***A review of a prospective data base for pediatric surgical admissions from January 2012 to December 2015 was made and examined all the entries for children admitted with biliary atresia.***
Results: ***In this study 46 patients were recruited with an age range at admission of 2 weeks to 3.5 years and a peak age of 2 months. During the four years, 14 Patients had portoenterostomy done and of these 5 died within 7 days after surgery. Thirty two (32) patients were not operated, 18 of them died and 13 were still alive by the close of 2015.***
Conclusion: ***A big number of children with biliary atresia presented late with decompensated liver functions having lost time in peripheral health facilities being managed for medical jaundice.***

Key words: Biliary atresia, Uganda, Jaundice.
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Introduction

Biliary atresia is characterized by biliary obstruction of un known origin and is the commonest cause of cholestatic jaundice in neonates due to inflammatory damage to the intra and extra hepatic bile ducts with sclerosis , narrowing and obstruction[1]. It has an incidence of 1:15000 live births, and has two clinical phenotypes; the syndromic or embryonic form that accounts for 10-20%, associated with congenital anomalies like polysplenia/asplenia, situs inversus, cardiac defects, absence of inferior vena cava and preduodenal portal vein, the perinatal or acquired/isolated form that accounts for 80-90% [2, 3]. Biliary atresia commonly presents with jaundice, acholic stools / dark urine and hepatomegaly but may be associated with complicationsdepending on the extent of the disease [3, 4, 5]. This disease rapidly leads to liver cirrhosis and liver failure if untreated therefore a timely Kasai- portoenterostomy restores bile flow enhancing survival without liver transplant and thus age at diagnosis of biliary atresia is a potentially modifiable risk factor [6]. We present data from a referral center analyzing the epidemiology and outcome of children admitted with biliary atresia in a period of four years. This study was aimed at establishing the epidemiology of patients presenting with biliary atresia and immediate surgical outcome.

Methods

A review of a prospective data base maintained for pediatric surgical admissions for 4 years from January 2012 to December 2015 was made and examined all the entries for children admitted with a diagnosis of biliary atresia. Included all the children who presented with jaundice and pale stools. The following parameters were evaluated; age at diagnosis /admission, sex, duration of symptoms, distance to access care, surgery, complications at admission and final outcome. Permission to study and publish the information was obtained from the hospital ethics and research review board.

Results

In a period of 4 years, 46 patients with confirmed biliary atresia were admitted at Mulago National Referral Hospital -Pediatric Surgery Unit. Amongst these 24(52%)babies were males and 22 (48%) were females.The age range at admission was 2 weeks to 3.5 years of age and a peak age of presentations was 2 months (see Table 1).

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During the four years, 14 patients were operated (Kasai's portoenterostomy) and of these 5 patients died within 7 days after surgery. Thirty two (32) patients were not operated, 18 of them died and 13 were still alive by the close of 2015. Average distance travelled by patients for care ranged from 5 km to 800 km from neighboring South Sudan. The longest duration was one patient who had jaundice and pale stools for 9 months. At presentation 21 patients had complications and 25 had no complications.

Table 1. Distribution of patients by age at presentation

Age at presentation	Number
2 weeks	3
3 weeks	3
1 months	2
2 months	10
3 months	4
4 months	5
5 months	4
5 months	4
6 months	8
7 months	3
8 months	2
9 months	2
2.5 years	1
3.5 years	1
Total	**46**

Table 2. Patient Distribution by Mode Of Admission

Mode of referral	Number	Percentages
Routine outpatient clinics	9	20%
Through emergency unit	11	24%
Came in as referrals from other hospitals	26	56%
Total	**46**	

Table 3. Distribution of Patients by Duration of Symptoms

Duration of symptoms (jaundice)	Number	Percentages
<1 month	17	37%
> 1mont h	29	63%
Total	46	

Table 4. Distribution by Type of Complications

Complications	Number
Failure to thrive (FTT)	7
Septicemia	2
Ascites	8
Liver cirrhosis	3
Portal hypertension	1
Total	**21**

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Discussion

In a span of 4 years 46 patients with biliary atresia were admitted averaging to 11.5 cases admitted per year compared to a study conducted in Brazil that reported a range of 1-13 cases per year with an average of 4.5 cases per year [7]. Another study conducted at the Children's Hospital and Medical center, Seattle Washington between 1989 to 1993 recorded 23 cases in 4 years [8]. In our study 24(52%) babies were males and 22 (48%) were females compared to other studies that have significantly reported more females than males.

The age range at admission was 2 weeks to 3.5 years of age and a peak age of presentations was 2 months which signifies that most babies present late to the center and its reflected in the low numbers of patients who had a portoenterostomy done (14/46) because the rest of the patients were considered to be of advanced disease at admission yet there are no liver transplantfacilities. A study in Seattle Washington reported a median age of referral for biliary atresia to be 61 days with a mean of 51 days which was rather late attributed to delays in investigating neonatal jaundice and acholic stools and this declines the success of portoenterostomy [8, 9]. This sharply contrasts the situation in the industrial world where the median age at diagnosis is 40 days and median age at Kasai operation is between 54 and 69 days of age [10].

Of notice 63% of patients presented with jaundice for more than 30 days this can be explained by the long distances travelled by patients to access the only center with specialized pediatric surgical services in the country but also compounded by the fact that patients get delayed in peripheral health facilities being treated for medical causes of jaundice as shown by 56% of admissions coming in as referrals from other hospitals. As results patients present with complicated disease i.e. 21/46 (45%) presented with one of the following complications at admission; failure to thrive, sepsis, ascites, liver cirrhosis, and portal hypertension. This reflects complete lack of screening strategy for biliary atresia as reported in some come countries by using colored (chromatic) cards with a stool color scale distributed to parents [6, 7, 10].

The mortality amongst operated children (5/14) 36% and the unoperated (18/32) 56% are still very high probably due to low expertise, lack of infrastructure and support services. Efforts must continue to be made to eliminate delayed diagnosis in order to improve success of Kasai and reduce the necessity of liver transplantation [10].

Conclusion

A big number of children with biliary atresia presented late and for the first time with decompensated liver functions having lost time in peripheral health facilities being managed for medical jaundice.

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