RESEARCH PAPER: SURGICAL MANAGEMENET OF BILIARY TREE DISEASES IN CHILDREN AT TIKUR ANBESSA SPECIALIZED HOSPITAL –A 10 YEAR RETROSPECTIVE STUDY.

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Declaration

I **Abraham Teshome** declare that this paper is a result of my independent research work on the topic entitled "***Surgical management of biliary tree diseases in children at Tikur Anbessa Specialized Hospital –a10 year retrospective study''*** in partial fulf illment of the requirements for specialty certificate for General Pediatric Surgery at Addis Ababa University, College of Health Sciences, Department of Surgery, Pediatric Surgery Unit. This work has not been submitted for a degree to any other university. All the references are also acknowledged.

Dr. Abraham Teshome

Signature: _____

Date: ___/_____/_____

Confirmation

This is to certify that Abraham Teshome has carried out this research work on the topic entitled "***Surgical management of biliary tree diseases in children at Tikur Anbessa Specialized Hospital –a 10 year retrospective study***" under my supervision. This work is original in nature and has not been presented for a degree in any University and it can be submitted for the partial fulfillment of the requirements for the award of the specialty certificate for General Pediatric Surgery.

Dr. Hanna Getachew (Assistant Professor of General & Pediatric Surgery)

Signature: _____

Date: _____/_____/_____

ACKNOWELEDEMENT

All glories go to Almighty God. Thank you Lord for giving me the strength and courage throughout my journey until now.

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Contents

Abbreviations

TASH	Tikur Anbessa Specialized hospital
IVC	Inferior Vena cava
BA	Biliary atresia
KP	Kasai Procedure
HJ	Hepaticojejunostomy
UDCA	ursodeoxycholic acid
U/S	Ultrasound
PI	primary investigator
RYHJ	roux-en-y-hepaticojejunostmy

OPERATIONAL DEFINITIONS

Kasai porto-enterostomy-surgical treatment performed for infants with biliary atresia to allow for bile drainage.

Triangular cord sign-a triangular or tubular echogenic cord of fibrous tissue seen in the porta hepatis at ultrasonography and is relatively specific for the diagnosis of biliary atresia.

Cholangitis- is an ascending bacterial infection in association with partial or complete obstruction of the bile ducts. The most common presentation is fever, epigastric or right upper quadrant pain, and jaundice.

Early outcome-post operative outcome in the 1st 6 months for patients for whom kasai Porto-enterostomy done.

Abstract

Background Information-biliary tree diseases are diseases affecting bile ducts, gallbladder and other structures involved in production and transportation of bile**.** A diverse spectrum of diseases affects the biliary system, often presenting with similar clinical signs and symptoms. These conditions include gallstones, cholecystitis, cholangitis, biliary tract cysts, congenital biliary atresia and others.

Biliary atresia is a progressive obstructive cholangiopathy of unknown etiology, occurring during the perinatal period. If left untreated it progresses to liver fibrosis and cirrhosis in the first few months of life. Timely Kasai porto-enterostomy restores bile flow enhancing survival and thus age at diagnosis is a potentially modifiable risk factor.

choledocal cyst is a rare congenital anomaly of the bile duct which is defined as pathological dilatation of the bile duct. If left untreated it has complications which range from biliary duct obstruction to cholangiocarcinoma. Total cyst excision and biliary reconstruction is the treatment of choice.

Little is known about the epidemiology of cholelithiasis in children. Cholelithiasis and choledocholithiasis have been increasingly diagnosed in recent years in children. This phenomenon may be attributed to better medical imaging (especially ultrasonography) and its usage in investigating children with unexplained abdominal pain and/or a genuine increase in the incidence.

Objective- to assess clinical presentation, clinical course and outcome of pediatric patients operated at TASH with biliary tree diseases.

Methodology: a retrospective study using structured questionnaire was used to collect data on pediatric patients operated at TASH for the diagnosis of biliary tree diseases from Jan 2010 to Jan 2020.The collected data was checked for completeness and analyzed using software SPSS 23.

Results and discussion-60% of patients operated for biliary tree and 87% of choledocal cysts were females. More than 60% of biliary atresia patients were operated within the 1^{st} 3 months.75% of choledocal cyst was type I and the rest type II.1/3 of patients operated for BA had their jaundice cleared on follow up.

Conclusion-age at KPE and post kasai jaundice clearance is low in our setting but similar with other African countries. Type II choledocal cyst is the 2^{nd} commonest type in our setting unlike other studies.

1.INTRODUCTION

1.1. BACKGROUND INFORMATION

Biliary tree diseases are diseases affecting bile ducts, gall bladder and other structures involved in production and transportation of bile. They are uncommon in childhood but should be included in the differential diagnosis of an infant or a child with jaundice, abdominal pain or an abdominal mass. Most of these diseases have no adult counterpart. And therefore should be approached differently.

Biliary atresia is a progressive obstructive cholangiopathy of unknown etiology, occurring during the perinatal period. If left untreated it progresses to liver fibrosis and cirrhosis in the first few months of life, with death occurring in the first few years (12). It is the leading cause of end-stage liver disease in the pediatric population and remains the most common indication for liver transplantation in children (13). The current surgical management of biliary atresia involves hepatic portoenterostomy (Kasai procedure) to re-establish bile drainage, thus delaying the progression of fibrosis, with subsequent liver transplantation still required in many cases.

Biliary atresia is the commonest cause of cholestatic jaundice in neonate due to inflammatory damage to the intra and extra hepatic bile ducts with sclerosis, narrowing and obstruction. It has an incidence of 1:10,000 -1:20000 live births (16) and has two phenotypes; the syndromic or embryonic form that accounts for 10-20% associated with congenital anomalies like polysplenia / asplenia, situs in versus, cardiac defects, absence of IVC, predeudnal portal vein and perinatal or acquired/isolated form which accounts for 80-90%(14). Biliary atresia commonly presents with jaundice, acholic stool/dark urine and hepatomegaly but may be associated with complications depending on the extent of the disease. Timely Kasai porto-enterostomy restores bile flow enhancing survival and thus age at diagnosis is a potentially modifiable risk factor.

Choledocal cyst is a rare congenital anomaly of the bile duct which is defined as pathological dilatation of the bile duct (15). If left untreated it has complications which range from biliary duct obstruction to cholangiocarcinoma. It is estimated that it has an overall incidence of 1 in 100000 to 1in 150000live births in western populations with a 3-4:1 female predominance,75% of which are diagnosed in childhood.

Total cyst excision and biliary reconstruction is the treatment of choice. Biliary reconstruction may be achieved by one of several techniques based on the surgeon's personal preference and the current circumstances of each case. Roux- en- Y hepaticojejunostomy (RYHJ) and to less extent hepatico-deudnostomy (HD) are commonly used, but many other procedures such as jejunal interposition hepatico-deudnostomy and appendix interposition have been reported. Stricture at the site of anastomosis is considered the main factor of repeated infection resulting in biliary stasis and stone formation.

Little is known about the epidemiology of cholelithiasis in children. Cholelithiasis and choledocholithiasis have been increasingly diagnosed in recent years in children. This phenomenon may be attributed to better medical imaging (especially ultrasonography) and its usage in investigating children with unexplained abdominal pain and/or a genuine increase in the incidence of cholelithiasis due to increasing use of total parenteral nutrition, frusemide and phototherapy in the infants (17). The exact prevalence of gallstones in children is not known. Studies from Europe have shown an overall prevalence of gallstone disease of 0.13% to 0.2% in children (18,19). The condition may present by right upper quadrant pain (75-85%), followed by nausea or vomiting (60%). Jaundice is less frequent and epigastric tenderness is present in one third of patients. cholelithiasis can be asymptomatic in 17% of children (21,22).

Biliary peritonitis from bile duct or gall bladder perforation is rare entity in children. In pediatrics CBD may be perforated spontaneously or due to anomalous pancreatico-biliary ductal system, congenital weakness of CBD, trauma, associated with choledocal cyst, viral infection, stenosis of CBD, iatrogenic or stone in CBD. The causes of gall bladder perforation include acalculous cholecystitis, trauma, or gall stones.

1.2. Statement of the problem

The etiology of BA is not known, likely multifactorial. These factors may include immune mediated ductal injury, viral agents as inflammatory triggers and predisposing genetic factors. The diagnosis of BA is sometimes challenging because of high degree of overlap in clinical, radiologic, and histologic characteristics with other causes of hepatitis in neonate. However, with combination of investigations it is possible to be reasonably certain in most cases. The current treatment of BA is surgical. Hepatoportoenterostomy for the relief of biliary obstruction in these infants was initially reported in 1959 by Kasai (8).

If possible, Kasai procedure should be performed before 60 days of age when its short term success is 80%(9,10,11). Efficacy of the procedure drops with the age of the patient, decreasing to 60% by 90 days. Although the prognosis is worse and the need for transplantation is higher in infants who undergo Kasai procedure after 3 months of age, most hepatologist feel the procedure should be attempted in most of these patients.

1.3. Significance of study

The objective of this study is to describe experience and outcomes of pediatric patients operated at Tikur Anbessa Specialized Hospital for biliary tree disease. The aim of this study was to identify studies that would be useful in comparing our data to other cohorts, in both developed and developing countries. There is little information available in an African context, and there was no study done in Ethiopia concerning surgical management of biliary tree diseases in pediatrics patients.

2.LITERATURE REVIEW

The study done in Red Cross War Memorial Children's Hospital in South Africa shows the median age at presentation in the 80 cases reviewed was 70 days. Kasai procedure (KP) was performed in 62 (77.5%) patients at a median age of 68 days. 18 patients who presented late did not undergo KP. Clearance of jaundice was achieved in 39% of KPs. 13 patients underwent KP beyond 90 days with a success rate of 38%. 2- and 5-year long term survival with native liver rates were 41% and 37.5% respectively with overall survival of 59% at 2-years and 56% at 5-years. Liver transplant was only performed in 12 of the 54 patients who showed progression to require transplantation. (1)

In a period of 4 years 46 patients with confirmed biliary atresia were admitted at Mulago National Referral Hospital-Pediatric surgery unit in Uganda. Amongst 24(52% were males and 22(48%) were females. The age range at admission was 2weeks to 3.5 years. During 4years 14 patients were operated (Kasai Porto - enterostomy) and of which 5 patients died within 5 years after surgey.32 patients were not operated ,18 died and the other 13 were alive by close 2015.The longest duration was a patient who had jaundice and pale stool for 9 months. At presentation 25 patients had complications. (**2)**

From the study done in three tertiary health facilities in South East Nigeria from January 2007-December 2011, a total of 31 consecutive cases of biliary atresia were seen. 24 cases of biliary atresia were included comprising 10 (41.7%)males and 14(58.3%) females. Seven (22.6%) were excluded due to incomplete data or lost follow up. The mean age of presentation was 4.02 months; range 1.75-11 months.15(62.5%) had surgery while 9 (37.5%) received medical treatment only. The mean age at death was 14.2months; range 2.5-30months. **(3)**

From cases diagnosed between March 1993 to February 1995 to pediatric surveillance unit in Britain 93 were confirmed cases of BA with a frequency of 1/16700 live births. Primary surgery was done in 91 children in 15 surgical centers with an early success rate for clearing jaundice of 55% overall. Centers were grouped according to caseload; group A had more than 5 cases/year and group B fewer than 5 cases/year. Early success was higher in group-A center, odds ratio 2·02 (95% CI 0·86–4·73), but this did not reach statistical significance. Of 41 children in whom surgery was unsuccessful in clearing jaundice 9 (22%) died and 30 (73%) underwent liver transplantation. Survival without liver transplantation and overall survival were both

significantly greater in group-A centers, rate ratios 0·48 (95% CI 0·27–0·86) and 0·32 (0·11–0·94). Actual 5-year survival without transplantation was 61·3% in group-A centers and 13·7% in Group-B centers. Actual 5-year overall survival was 91·2% in group A and 75% in group B. Once center size was taken into account, no other factor, including age at surgery, was predictive of survival without transplantation or overall survival. **(4)**

From a case series done at pediatric surgery department at Hospital civil de Guadalajara in Mexico they found a total of 14 patients with choledocal cyst from which 10 (71.2%) were females and the rest were males with the ratio of 2.5:1.The average age of presentation was 5.2 years (ranges from 1 to 15 years).The combination of abdominal pain and jaundice was the predominant symptomatology presented in 64.2% of their population .The classic triad of jaundice, abdominal pain and palpable mass was only present in 2 patients.2 of the patients had spontaneous perforation of the CC found as incidental finding during laparatomy.The surgical approach consisted of excision of the cyst in all patients with hepaticojejunostomy in 8(57.1%) and hepaticodeudnal anastomosis in 6(42.9%)(**5**)

In Egypt retrospective study included 27 patients with choledocal cysts, who were treated during the period from 1993 to 2005. Complete excision of the extra-hepatic cysts was done in all patients. The study population was classified into two groups according to the method used for biliary reconstruction. Group 1 included 18 patients who undergone RYHJ, while group 2 included 9 patients treated with HD. All patients were studied as regard to the details of clinical presentation, diagnostic tools, operative details, and outcome. Total excision of choledocal cyst is usually feasible. Both RYHJ and HD are effective techniques for biliary reconstruction following excision of the cyst with satisfactory and comparable results on both early and long-term follow up. Hepatico-deudnostomy may be preferred due to shorter operative time and avoidance of intestinal anastomosis; however, more patients with HD are required before reaching a solid conclusion. (**6**)

Review of 84 patients who had choledocal cyst cared for by the Division of Pediatric Surgery, Department of Surgery, The University of Hong Kong, Queen Mary Hospital over a period between 1965 and 1996 was undertaken. Of the 84 patients who had choledocal cyst who came under our care, 79 have had definitive surgery, three are awaiting surgery, one is being observed with Caroli's disease, and the parents of one child have refused surgery. The ages of our patients

ranged from 2 weeks to 14 years (mean age 3.5 years). Follow up of patients after surgery ranged from 4 months to 17 years. 41 patients have had cyst excision with hepaticojejunostomy using a 40-cm Roux loop without an anti- reflux procedure.

Early complications in those who underwent cyst excision with hepaticojejunostomy included anastomotic leak in three patients who required reoperation, cholangitis in two, and fluid collection in the gall-bladder bed that required no intervention in one. In the 41 patients who underwent cyst excision, there was no early or late mortality. There was no observed malignant transformation in any of our patients either before or after EX-HJ. Cyst excision was associated with anastomotic leak in our first three (7%) cases, and the sites of leakage were at the hepaticojejunostomy in two, and at the jejunojejunostomy in one.

Internal drainage procedures performed in the early years of this study, however, were associated with significant morbidity and mortality. Anastomotic leak occurred in three of 38 patients (8%), and the sites of leakage were at choledochorrhaphy in two and at cystojejunostomy Roux-en-Y in one. (7)

In retrospective study done from January 2005 to September 2010 in Central Anatolia, Turkey,124 children with sonographically diagnosed gallstone were stratified into group 1 (symptomatic) and group 2 (asymptomatic). The data on demographic features, predisposing risk factors, laboratory features, complications, and outcome were collected from medical charts and compared by using convenient statistical methods. There were 76 (61%) children in group 1. Females were significantly older than males at the time of diagnosis (p=0.001). After adjusting for age and sex, asymptomatic presentation was associated with hemolytic anemia (r=346, <0.001) and being an oncologic patient (r=248, p=0.006). No risk factor was specifically associated with having a symptomatic presentation. Sixteen children (12.9%) developed complications: 14 (18.4%) in group 1 and 2 (4.2%) in group 2 (p=0.027). Gallstone resolution was detected in 20 (29.4%) and 10 children (23.3%) in groups 1 and 2, respectively (p=0.477). Resolution was observed in 43.8% of children with ceftriaxone-associated gallstone. The rate of resolution with ursodeoxycholic acid (UDCA) was similar to that observed with expectant management. Gallstone resolution was evident in 9 infants (50.0%) and was significantly higher than children over 2 years of age (21 out of 106 children, 19.8%) (p=0.006). The most important factor associated with gallstone resolution was to be an infant (<2 years of age) at the time of

diagnosis. **Conclusion**: Ceftriaxone-associated gallstones are most likely to resolve but do not always undergo spontaneous resolution. UDCA treatment seems to be ineffective. Young age is a favorable factor for gallstone resolution. The rate of complications in children with asymptomatic presentation is considerably low. Thus, clinical follow-up rather than surgical intervention is suggested in children with asymptomatic presentation and in infants. (20)

From prospective study performed in Cairo university pediatric hospital between 2004-2009 on 30 pediatric patients,19 males and 11 females, their age ranged between 4 months and 14 years, the mean age was eleven years. Fifteen cases were with hemolytic disorder. The diagnosis of cholelithiasis was made by ultrasonographic examination in all cases. Preoperative management included correction of anemia (2 cases with sickle cell with severe anemia were given blood transfusion and were operated upon after improvement of their blood profile). Cases with cholangitis (one case) or calcular pancreatitis were treated and postponed till subsidence of the attack. Laparoscopic cholecystectomy was performed to 20 patients and open Cholecystectomy to 8 patients and 2 patients were followed up for 6 months on ursodeoxycholic acid and repeated U\S every 2 months till their U\S revealed gall bladder free of stones and those 2 cases were below one year and they are still on follow up. No major biliary leaks or injury to common bile duct and no mortality. Post-operative hospital stay ranged from 2 days to 7 days with mean of 3 days

Conclusion- Biliary stones disease has become more common in children and is often idiopathic, asymptomatic children with non-calcified gallstones and without associated comorbidities can be managed expectantly especially in the very young population (below one year). Symptomatic gallstones should be managed promptly especially in the hemolytic patients. Laparoscopic cholecystectomy is safe and effective in the pediatric age and should be into the practice of pediatric surgeons. (23)

3.OBJECTIVES

3.1. General objective

To assess clinical presentation, clinical course and outcome of children operated at TASH with biliary tree diseases.

3.2. Specific Objectives

1.to assess clinical presentation of children with biliary tree diseases operated at TASH.

2.to assess clinical course of children with biliary tree diseases.

3.To assess outcomes of children operated for biliary tree disease.

4. Methods and Materials

4.1. Study Area

The study was conducted in TASH, Addis Ababa City, in Ethiopia. TASH is a tertiary level referral and teaching hospital which deliver comprehensive care to pediatric patients including to those with biliary tree surgical problems.

4.2. Study period

From January 1, 2010 up to January 30, 2020

4.3. Study design-

Retrospective descriptive study.

4.4. Source population

All children operated at TASH from January 1,2010 to January 30,2020 on elective basis at pediatric surgery unit.

4.5. Study population

All children operated at TASH for the diagnosis of biliary tree diseases.

Inclusion criteria-children operated for biliary tree diseases over the specified period.

Exclusion criteria-lost documentation and charts and patients operated on emergency bases.

4.6. Data collection procedures and analysis

Data was collected with standard questionnaire prepared by the investigator from the patient's chart.

The collected data was checked for completeness and data analysis was done by manual and computer using software (SPSS).

4.7. Study Variables

Independent variables-age, sex, address, jaundice, Pale stool, Organomegally.

Dependent-type of procedure, anastomotic leak, peri-operative death, jaundice clearance.

4.8. Ethical Issue

Ethical letter was written from the surgical department for card recruitment from the card room. The research proposal was approved at department research and publication committee.

RESULTS AND DISCUSSIONS

RESULTS

Part I- Demography of patients

Table 1-Gender of patients operated for biliary tree diseases

Gender	frequency	Percent
Male	10(all)	40
	1(choledocal cyst)	12.5
Female	15(all)	60
	7(choledocal cyst)	87.5
Total	25	100

60%of the operated patients for biliary tree diseases were females overall, but females account for more than 87 % of cases operated for choledocal cyst.

Table 2-address of the patients

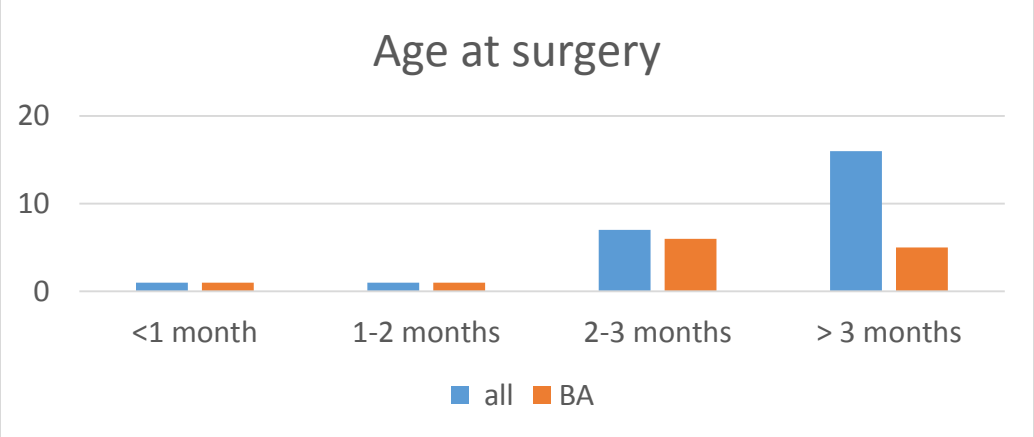
address	Frequency	Percent
Addis Ababa	11	44
Outside Addis	14	56
Total	25	100

56% of the patients came from outside Addis.

Table 3-Cross tabulation of the patients addresses with age at presentation

			age of the patient				
			< 1 month	1-2months	2-3monts	>4months	Total
address of the **patient**	Addis Ababa	Count	1	0	1	4	6
		% within address of the patient	16.7%	0.0%	16.7%	66.7%	100.0%
	Outside Addis	Count	0	1	5	1	7
		% within address of the patient	0.0%	14.3%	71.4%	14.3%	100.0%
Total		Count	1	1	6	5	13
		% within address of the patient	7.7%	7.7%	46.2%	38.5%	100.0%

Figure 1-AGE DISTRIBUTION OF PATIENTS



Overall 16 patients (65%) were operated after 3 months of age. But more than 61% of the patients with biliary atresia were operated within 3 months of age.

Part II-clinical presentations and course

Table 4-duration of symptoms

64 % of the operated patients were symptomatic for more than a month

Table 5- Level of direct bilirubin pre-operatively

Duration	**Frequency**	**Percent**
< 2weeks	3	12.0
2-4 weeks	6	24
>4 weeks	16	64
Total	25	100.0

bilirubin	Frequency	Percent
<2	6	24
2-10	12	48
>10	5	20
Unknown	2	8
Total	25	100

Significant number of the patients (48 %) had direct hyperbilirubinemia in the range of 2-10 mg/dl.

Table 6-hepatosplenomegally

Hepatosplenomegaly	Frequency	Percent
Yes	13	52
No	12	48
Total	25	100

52% of patients had hepatosplenomegaly at presentation

Table 7-clinical presentation in BA patients

	Jaundice	**Pale stool**	**Dark urine**
Yes	13(100%)	11(84.6%)	8(61.5%)
No	0	2(15.4%)	2(23.1%)
Not explained	0	0	3(15.4%)
Total	13(100%)	13(100%)	13(100%)

84.6% of patients operated for biliary atresia had pale stool on presentation.61.5% had dark urine on presentation.

50% of patients diagnosed with choledocal cyst are jaundiced and only 1 patient had triad of jaundice, pain and abdominal mass. There was no patient with choledocal cyst diagnosed intra uterine

Table 8-Type of imaging and imaging findings

		Frequency	Percent
Type of imaging(for choledocal cyst)	Ultrasound only	4	50
	CT scan	1	12.5
	Ultrasound+ CT/MRI	3	37.5
Gall bladder on imaging	Normal size	3	12.0
	Small/rudimentary	5	27.0
	not visualized	3	12.0
	not applicable	14	56.0
Triangular cord sign	Yes	8	32
	No	3	12
	Not applicable	14	56
	Total	25	100.0

50% of the patients with choledocal cyst had ultrasound only while 37.5% had a combination of ultrasound and CT or MRI.3 patients (23%) with biliary atresia were imaged with CT/MRI in addition to U/S. Triangular cord sign was seen on 8 patients out of 11 patients diagnosed with BA

Table 9-Choledocal cyst-types

Type of choledocal cyst	Frequency	Percent
type I	6	75
type II	2	25

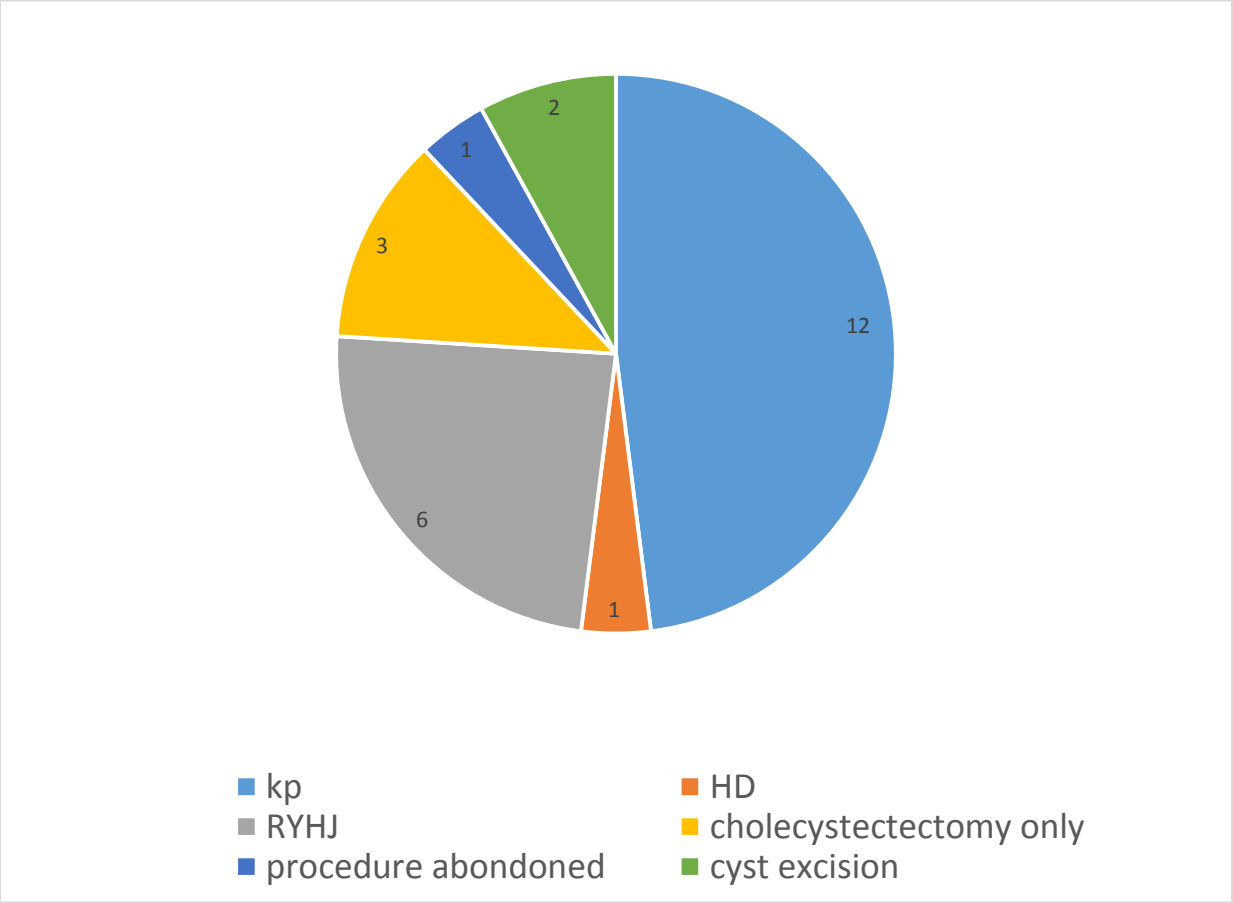
The commonest type of choledocal cyst was Type I which accounts 75% followed by type II and no other types seen.

Table 10-Diagnosis

Pre-operative	Diagnosis	Frequency	Percent
	biliary atresia	11	44.0
	choledocal cyst	10	40.0
	cholelithiasis/choledocholithiasis	3	12.0
	Others	1	4.0
Intra-operative	Biliary atresia	13	52.0
	Choledocal cyst	8	32.0
	Cholelithiasis/choledocholithiasis	3	12.0
	Others	1	4.0
	Total	25	100.0

The most common disease operated was BA followed by choledocal cyst.2 of the patients pre-operatively diagnosed as choledocal cyst became BA intra-operatively.

Figure 2- Type of procedure done



The dominant procedure was KP which was done for 12 patients followed by RYHJ. two patients had cyst excision for type II choledocal cyst.one patient with CBD stricture undergone hepatico-duodenostomy.one of the patient with BA had exploration and procedure deferred because it was advanced disease.

There was no patient for whom liver biopsy done and for whom intra-op cholangiography done.

Part III-post operative course

Table 11-post op prophylactic antibiotics & steroid for BA patients

	Antibiotics	Steroid
Yes	6(46.2%)	3(23.1%)
No	4(30.8%)	7(53.8%)
Not applicable	3(23.1%)	3(23.1%)
Total	13(100%)	13(100%)

46% of patients were given prophylactic antibiotics and 53.8% of patients were not given steroid post kasai porto-enterostomy at discharge.

Table 12- Post –op conditions

		Frequency	Percent
Complications	Yes	3	12.0
	No	22	88.0
Type of complication	Anastomotic leak	1	4.0
	HAI	2	8.0
	Not applicable	22	88.0
Post op	Discharged improve	22	88.0
	Discharged the same	1	4.0
	Died	2	8.0
Cause of deaths	HAI	1	4.0
	Anastomotic leak/GI onset sepsis	1	4.0
	Not applicable	23	92.0
	Total	25	100.0

12 % of the operated patients had post op complication.2 of the patients had HAI ,1 patient had anastomotic leak.2 of the 25 operated cases died post operatively.88% of the patients discharged improved.

Table 13- Post-operative follow up

		Frequency	Percent
Jaundice clearing(for BA)	Yes	4	30.7
	No	4	30.7
	not applicable	5	38.6
Re-admission for cholangitis	Yes	4	16.0
	No	19	76.0
	not applicable	2	8.0
Re-explored	Yes	2	8.0
	No	22	88.0
	not applicable	1	4.0
Indication for re-exploration	post op leak	1	4.0
	bowel obstruction from adhesion	1	4.0
	not applicable	23	92.0
	Total	25	100.0

From patients operated for biliary atresia only 30.7% had their jaundice clearing .4 patients were re-admitted for post op cholangitis.2 of the operated cases were re-explored,1 for post op adhesive bowel obstruction and the other for post KP anastomotic leak

Table 14-current status of the patient operated for BA and choledocal cyst

Diagnosis	Alive	Died	Phone not working
BA	1	7	4
Choledocal cyst	5	1	2
Total	6	8	6

-currently 1 patient is alive from 12 post kasai patients. Only 1 patient died post RYHJ for choledocal cyst. There was no a single patient for whom liver transplantation done post kasai Porto enterostomy.

DISCUSSION

Biliary tree diseases are diseases affecting bile ducts, gall bladder and other structures involved in production and transportation of bile. They are uncommon in childhood but should be included in the differential diagnosis of an infant or a child with jaundice, abdominal pain or an abdominal mass.

More than **61%** of the patients with biliary atresia were operated **within the 1st 3 months** of age. This is similar with study done in Red Cross War Memorial Children's Hospital in South Africa showing the median age at presentation in the 80 cases reviewed was 70 days. Kasai procedure (KP) was performed in 62 (77.5%) patients at a median age of 68 days.

Only **one third (30.7 %)** of biliary atresia patients had their jaundice **clearing** post operatively on follow up. This is lower than the study done in Red Cross War Memorial Children's Hospital in South Africa in which Clearance of jaundice was achieved in 39% of KPs. But it is higher than the result found in the study done in30‐month period between January 2009 and June 2012 at two academic hospitals in Johannesburg; of the 43 children undergoing KPE, 12 (27.9%) achieved early resolution of jaundice, while 6 (14.0%) patients were already lost to follow‐up by 6 months, and 7 (16.3%) had died.

Jaundice clearance in our setting is significantly lower than that found in cases diagnosed between March 1993 to February 1995 to pediatric surveillance unit in Britain 93 were confirmed cases of BA with a frequency of 1/16700 live births. Primary surgery was done in 91 children in 15 surgical centers with an early success rate for clearing jaundice of 55% overall.

In our study,**2** of the 13 patients operated for biliary atresia died post operatively accounting for **15%**,1 from HAI and the other from anastomotic leak. In retrospective analysis of all children who underwent KPE in a 30‐month period between January 2009 and June 2012 at two academic hospitals in Johannesburg was undertaken, Of the 43 children undergoing KPE, 7 (16.3%) had died. The 7 early deaths included 1 child with postoperative complications and nosocomial pneumonia, 1 with upper gastrointestinal bleeding and suspected sepsis, 1 with HIV World Health Organization stage‐IV infection and chronic diarrhea, and 1 with septicemia. The cause of death for the other 3 was unknown.

Mortality in our setting is lower than that found in prospective review done from 2012 to 2015 in Makerere university of Uganda in which 7 days' post-surgery mortality was 36%(5 of 14) operated patients.

50% of patients diagnosed with choledocal cyst are **jaundiced** and only **1** patient had **triad** of jaundice, pain and abdominal mass which is similar with the study done at Hospital civil de Guadalajara in Mexico where they found a total of 14 patients with choledocal cyst in which classic triad of jaundice, abdominal pain and palpable mass was only present in 2 patients. The **sex ratio** of patients with choledocal cyst was **7F:1M** but in the study stated above 10 (71.2%) were females and the rest were males with the ratio of 2.5:1 unlike in our case.

50% of the patients with choledocal cyst had ultrasound only while 37.5% had a combination of ultrasound and CT or MRI. In Egypt retrospective study included 27 patients with choledocal cysts, who were treated during the period from 1993 to 2005 abdominal ultrasonography was performed in all patients. CT scan was done in only 8 patients, and MRI was performed in another 6 patients.

6 patients with choledocal cysts undergone **RYHJ and 2 patients had simple cyst excision only for type II cysts** in our set up. The commonest type of choledocal cyst was Type I which accounts 75% followed by type II and no other types seen. But in the study done in Tanta hospital in Cairo Twenty patients (74.1%) had type I; seven (25.9%) had type IV; while none had type II, III or V.

Conclusion

-age of kasai porto-enterostomy in our setting is similar with some of other African countries

-jaundice clearance post KP is low but similar with some studies in Africa

-post kasai peri-operative mortality in our setting was lower than some developing countries but similar with some of them.

-type II choledocal cyst is the 2nd commonest type of choledocal cyst in our setting unlike what is written in text books and the studies done in other countries in which this type is one of the rarest type,

-we are not practicing hepatico-deudnostomy commonly

-there was no patient for whom liver transplant done.

Limitations

-small sample size

-retrospective study

Recommendations

Farther study on outcome of patients treated medically for BA and compare long term outcome to those surgically treated.

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