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Original Research Article

Clinical study and management of surgical problem of hepatobiliary and pancreatic disease in children

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ABSTRACT

Background: Conditions which can be treated by surgery like biliary atresia etc., can lead to infantile jaundice. For better outcome it is important that the diagnosis be made quickly and precisely. There is danger of irreversible damage to the liver if prompt action is not taken. Here comes the role of diagnostic imaging which can help for prompt and precise diagnosis. This helps to act rapidly for the surgeon. The objective was to study the surgical problem of hepatobiliary and pancreatic disease in children.
Methods: Hospital-based follow up study was carried out among 10 pediatric patients with surgical problems hepatobiliary and pancreatic diseases. After admission detailed history of all the cases were taken according to the proforma approved by the guide information regarding the age at presentation to hospital, gestational age, birth weight, age at onset of jaundice, nature of symptoms, duration of symptoms was obtained.
Results: Out of the total cases, 50% constituted the biliary atresia cases followed by choledochal cyst in 30% of the cases. Chronic pancreatitis constituted 20% of the cases. In 20% of the patients the birth weight was less than two kg. All 5 patients had icterus. 3 patients had icterus. Ultrasound showed that the most common finding was hepatomegaly in 60% of the cases. All 5 patients had good uptake of radiotracer and excretion of radiotracer in intestine was poor in all 5 patients. The mean value of total bilirubin is 13.9.
Conclusions: Early and rapid intervention in biliary atresia cases reduces morbidity and mortality.

Keywords: Biliary atresia, Children, Hepatobiliary, Pancreatic disease

INTRODUCTION

Conditions which can be treated by surgery like biliary atresia etc., can lead to infantile jaundice. For better outcome it is important that the diagnosis be made quickly and precisely. There is danger of irreversible damage to the liver if prompt action is not taken. Here comes the role of diagnostic imaging which can help for prompt and precise diagnosis. This helps to act rapidly for the surgeon.[1] It has been said that the most common disorders affecting the digestive system are the pancreatic and hepatobiliary disorders. During embryonic development, hepato-pancreatobiliary system undergoes development of complex nature. Because of this complex nature, the chances of anomalies are more. This can affect the normal functioning of the system. It may also be associated with a number of other problems which are acquired. Highly skilled hand is required to deal with such problems. In this connection, it has been said that the disease of the gallstone is too much prevalent. It puts a lot of burden on the health care system. Multiple factors play their role in the process of the pathogenesis. It has been estimated that its prevalence is increasing now a days due to changes in the dietary habits, changes in the physical activity and life style changes.[2]

People are becoming more and more aware about the diseases and with the availability of the new diagnostic modalities and increase in the demand for such investigations by the patients is contributing to the diagnosis rapidly. It is also useful for the surgeons to manage the cases easily. There is more and more demand as well as more and more availability of skilled laparoscopic surgeons thereby making the number of laparoscopic hepatobiliary surgeries being performed more and more. Malignancy of the gall bladder is ranking fifth place amongst all the malignancies. Among the cancer of the biliary tract, it is the most common malignancy. It is aggressive in nature. Case fatality rate is very high. Natural history of the disease is not properly understood. Causes are also not well established. It has been estimated that it has a very high incidence in India. The cause has been attributed to presence of mineral in the waters of the north India. Surgery is the only treatment of choice in early stages but as the patients present in the last stages of the disease, the option of surgery stands at bay.[3]

With this background present study was carried out to study the management of surgical problem of hepatobiliary and pancreatic disease in children.

METHODS

This was a hospital based follow up study carried out at Pediatric Surgery Unit of Department of Surgery, Karnataka Institute of Medical Sciences, Hubli, Karnataka, India from November 2010 to October 2012.

During the study period, it was possible to study 10 pediatric patients with surgical problems hepatobiliary and pancreatic diseases. As this disease entity is rare in the children and it is difficult to get more cases at a single centre, author could include 10 cases in this study.

Patients with age less than 13 years and having surgical problems hepatobiliary and pancreatic diseases were included. Patients of age more than 13 years, having hepatobiliary or pancreatic lesions due to trauma and parents not willing to include their children in the present study were excluded.

Procedure

In cases of biliary atresia infants having pathological jaundice were admitted for evaluation of jaundice to rule out surgical causes of jaundice in infancy. After admission detailed history of all the cases were taken according to the proforma approved by the guide information regarding the age at presentation to hospital, gestational age, birth weight, age at onset of jaundice, nature of symptoms, duration of symptoms was obtained. All the patients underwent detailed examination, all patients had hemogram, blood urea, serum creatinine, and LFT tests were done. Ultrasound of the abdomen was done in all the patients, HIDA scan was done in all the 5 patients, prior to HIDA scan patients were treated with Phenobarbital (5mg/kg/day) for 3-5 and delayed images until 24 hrs were obtained. Based on HIDA scan findings i.e. failure to see the dye in intestine even after 24 hrs diagnosis of biliary atresia was made and, in this study, dye was not seen in the intestine even after 24 hrs in all the 5 patients. About 3 out of 5 biliary atresia patients underwent Kasai procedure and in remaining 2 patients Kasai procedure was not done because patients presented to hospital after 60 days of life and the need for liver transplantation was not available in this hospital patients were referred to higher centre for further management. Remaining 3 patients were posted for surgery, abdomen was opened through right sub costal incision, entire EHBA was atretic in all the 3 patients so author didn't go ahead with operative cholangiogram, directly author performed Kasai procedure in all the 3 patients and abdomen was closed by placing a drain. All the patients were given I.V. fluids, Nasogastric aspiration was done, and antibiotics and analgesics were given. Drainage tube was removed between 3-10 days based upon the drainage. In the present study, out of 3 biliary atresia patients 2 patients HPE revealed giant cell hepatitis and remaining 1 patient HPE showed intrahepatic cholestasis.

In choledochal cyst group, patients were admitted and evaluated for chronic pain abdomen. Detailed examination of the patient was done, investigations like hemogram, renal and liver function tests were done. Ultrasound abdomen was done in all 3 patients who revealed choledochal cyst, later MRCP was done in all 3 patients to confirm the diagnosis which revealed type 1 cyst in 1 patient and type IV A cyst in 2 patients. All 3 patients were posted for surgery, abdomen was opened through right sub costal incision and the choledochal cyst was identified, excision of the cyst was done with hepatic duodenostomy and abdomen was closed after placing a tube drain. All the patients were given I.V. fluids, Nasogastric aspiration was done and antibiotics and analgesics were given. Drainage tube was removed between 3 to 10 days based upon the drainage. In the present study, all the 3 patients histopathological examination report were consistent with choledochal cyst. There was no problem in the follow up period in any patient. Nothing more can be stated because of limited period of follow up of patients.

In chronic pancreatitis, group 2 patients were admitted with history of recurrent abdominal colicky type of pain over epigastrium with radiation of pain to back, after admission detailed examination of the patient was done, investigation like hemogram, blood urea, serum creatinine, liver function tests and serum amylase levels were done.

Patients were subjected to USG abdomen and later CT-scan of the abdomen was done, USG and CT- scan revealed chronic calcific pancreatitis with intraductal calcification with dilated pancreatic duct more than 6mm in both patients and in 1 patient there was also pseudo

cyst formation. Indication for surgery in both patients was multiple intraductal calculi in the pancreas with dilated pancreatic duct more than 6mm in both patients. Both patients underwent lateral pancreatico-jejunostomy and withstood the procedure well. There was no problem in the follow up period in any patient. Nothing more can be stated because of limited period of follow up of patients.

The data was entered in the Microsoft Excel sheet for the present study and analyzed using proportions.

RESULTS

Table 1 shows classification of patients based on disease group. Out of the total cases, 50% constituted the biliary atresia cases followed by choledochal cyst in 30% of the cases.

Table 1: Classification of patients based on disease group.

Disease group	No. of patients	%
Biliary atresia	5	50
Choledochal cyst	3	30
Chronic pancreatitis	2	20
Total	10	100

Chronic pancreatitis constituted 20% of the cases. All three patients having choledochal cyst presented after newborn period with pain, all were females, two had pain and vomiting, two had type 4A as per the anatomical classification and all of them underwent excision of the cyst with biliary enteric anastomosis (hepatico-duodenostomy).

On follow up there was no problem. Out of two cases with chronic pancreatitis, the cause could not be established, the age at presentation in these two cases was 5-10 years, all of them presented with abdominal pain, epigastric pain, radiating pain, colicky pain and nausea/vomiting, all of them had symptoms for more than two years before they presented to us, the amylase value was <200IU in both the patients, both of them could be diagnosed using ultrasound and CT scan, one of the patient had pseudo cyst, both the patients underwent lateral pancreatico-jejunostomy.

The five patients having biliary atresia are described here from Table 2 onwards.

Table 2: Distribution of patients by birth weight in biliary atresia.

Birth weight (kg)	No. of patients	%
<2	1	20
2-2.5	3	60
2.5-3	1	20
Total	5	100

Table 2 shows distribution of patients by birth weight in biliary atresia. In 20% of the patients, the birth weight was less than two kg while in 60% of the cases the birth weight was 2-2.5kg. Only 20% had normal birth weight.

Table 3: Distribution of patients by signs.

Signs	No. of patients	%
Icterus	5	100
Hepatomegaly	3	60
Splenomegaly	2	40
Ascites	0	0

Table 3 shows distribution of patients by signs. In the present study out of 5 patients in biliary atresia group, all 5 patients had icterus, 3 patients had hepatomegaly and 2 patients had splenomegaly.

Table 4: Distribution of patients by combination of signs.

Signs	No. of patients	%
Icterus with hepatomegaly	3	60
Icterus with splenomegaly	2	40
Total	5	100

Table 4 shows distribution of patients by combination of signs. In the present study, out of 5 patients in biliary atresia group, 3 patients had icterus with hepatomegaly, 2 patients had icterus with splenomegaly.

Table 5: Distribution of patients by ultrasound findings.

Ultrasound findings	No. of patients	%
Hepatomegaly	3	60
Splenomegaly	2	40
Visualized gall bladder	1	20
Triangular cord sign	1	10

Table 5 shows distribution of patients by ultrasound findings. Ultrasound showed that the most common finding was hepatomegaly in 60% of the cases followed by splenomegaly in 40% of the cases. One patient had triangular cord sign.

Table 6: Distribution of patients by HIDA scan findings.

HIDA scan findings	Uptake of radiotracer	Excretion of radio-tracer in intestine
Good	5	0
Poor	0	5

Table 6 shows distribution of patients by HIDA Scan findings. In the present study, out of 5 patients in biliary atresia group all 5 patients had good uptake of radiotracer and excretion of radiotracer in intestine was poor in all 5 patients.

Table 7: Distribution of patients of biliary atresia by LFT.

Test	No. of patients	Mean
Total bilirubin	5	13.9
Direct bilirubin	5	6.2
ALT	5	202.6
AST	5	324.4
ALP	5	415.2

Table 7 shows distribution of patients of biliary atresia by LFT. In the present study, in biliary atresia group the mean value of total bilirubin was 13.9, direct bilirubin mean value was 6.2, ALT enzyme mean value was 202.6, AST mean value was 324.4 and ALP mean value was 415.2.

DISCUSSION

In the present study, out of 5 patients in biliary atresia group 2 patients presented at 30 days of life, another 2 patients at 60 days of life and 1 patient at 90 day of life. Similar findings were reported by Matthai J et al, Yachha SK et al, Urganci N et al, noted that the average age of the patients in their study was 60 days.[4-6]

In the present study, out of 5 patients in biliary atresia group 3 patients were male and 2 were female. Dehghani SM et al, noted in their study that males were more than females. Donia AES et al, found that males and females were equally distributed in their study. Dick MC et al, and Wongsawaadi L et al, observed in their study that females were more than males.[7-10]

In the present study, out of 5 patients in biliary atresia group 1 patient gestation age was <32 week remaining 4 patients gestation age was between 38-40 weeks. Similar results were observed in way Lee WS studies where in median gestational age for BA was 40 weeks, in Urganci N et al, study all 14 patients in biliary atresia group were born at term.[6,11] In Porter CA et al, study out of 32 patients in EHBA group one child with EHBA had a gestation of < 37 weeks.[12] From the above studies and from the present study author concluded that patients with biliary atresia are born at normal gestational age.

In the present study, out of 5 patients in biliary atresia group 1 patient birth weight was <2kg, 3 patients birth weight was between 2-2.5kg and remaining 1 patient birth weight was between 2.5-3kg. Mean birth weight of patients with BA was 2951±556g in Dehghani SM et al, study. In Porter CA et al, study out of 32 patients none of these infants with EHBA weighed less than 2.5g at birth.[12]

Author found that all children with biliary atresia had jaundice. In Dehghani SM et al, study prevalence of alcoholic stools was 94.7% for BA.[7] In Urganci N et al, study jaundice was observed in 100% cases and alcoholic stools in 92.4% cases.[6] Thus jaundice and passing clay colored stools were most common presentation.

Author found that splenomegaly was present in 40% of the patients. In Urganci N et al, study hepatomegaly was present in 100% of cases where splenomegaly was present in 78.5% of cases.[6] Because of small sample size the results didn't match with above mentioned studies. Nonetheless hepatomegaly and splenomegaly are invariably present in biliary atresia cases with delayed presentation.

Using ultrasound to identify a normal gallbladder (>1.5cm in length), the infant must be fasting for 4 hours before the examination. The findings of a choledochal cyst or intrahepatic ductal dilatation effectively exclude the diagnosis of biliary atresia. If the gallbladder was shrunken or not visualized, biliary atresia was suspected.

A contraction in gallbladder size after feeding eliminates the possibility of biliary atresia. The fibrous cord remnant and portal plate found in biliary atresia can sometimes be visualized with sophisticated ultrasound techniques. The presence of a preduodenal portal vein or asplenia/polysplenia was further supportive of biliary atresia. Care must be taken to not misinterpret the parallel hepatic artery channel as a patient ductal structure. Hepatomegaly was revealed in 3 patients (60%), splenomegaly was found in 2 patients (40%) and visualized gall bladder was found in 1 patient (20%). In case of Urganci N et al, study hepatomegaly was found in 100% of cases and splenomegaly was found in 78.5% of cases.[6] Because of small simple size in this study, the results were not comparable.

In the present study, HIDA-scan was done in all the 5 patients of biliary atresia group. Failure to excrete radiotracer in to the intestine during HIDA-scan occurred in all 5 patients of biliary atresia (100%). Similar results were observed by Park WH and Kim MJ et al.[13,14]

In the present study, 3 out of 5 patients in biliary atresia group underwent Kasai procedure and in remaining 2 patients, Kasai procedure was not done because patients presented to hospital after 60 days of life and the need for liver transplantation was explained to be patient attenders as facilities of liver transplantation was not available in this hospital patients were referred to higher hospital. The incision used in Kasai procedure was right. Sub costal incision, EHBA was completely atretic in all 3 patients intraoperatively so didn't go ahead with operative cholangiogram. Portahepatis was anastomosed to the Roux-en-Y loop of jejunum. Abdomen closed by placing abdominal drain.

All the patients were given I.V. fluids, nasogastric aspiration was done and antibiotics and analgesics were given. Drainage tube was removed between 3 to 10 days based up on the drainage.

In the present study, out of 3 patients that underwent surgery 1 patient developed surgical site infection and burst abdomen and remaining patients didn't have any post-operative complication.

In the present study, out of 3 biliary atresia patients 2 patients HPE revealed giant cell hepatitis and remaining 1 patient HPE showed intrahepatic cholestasis.

CONCLUSION

Early and rapid intervention in biliary atresia cases reduces morbidity and mortality. Early diagnosis of the biliary atresia is possible if the ultrasound of the abdomen is done at an early stage of symptoms among the children.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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