SURGICAL BILIARY TREE PATHOLOGIES IN PAEDIATRIC PATIENTS

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ABSTRACT

Objective — *To find out various pathologies affecting biliary tree in paediatric age group and mode of investigations performed to arrive at diagnosis.*

Study design — *Descriptive case series.*

Place & Duration of study — *Department of Paediatric Surgey Unit B, National Institute of Child Health Karachi, from January 2007 to December 2007.*

Methodology — *All paediatric patients below the age of 12 years who presented with symptoms related to biliary tree were included. A detailed history was taken including presence of jaundice, color of stool, mass and pain in right upper abdomen etc. All wee subjected to ultrasound as a screening test. Laboratory investigations included blood complete picture, liver function tests including bleeding profile, TORCH titre where appropriate, HIDA scan and CT scan as indicated. Laparoscopy was done in cases with persistent jaundice for operative cholangiogram and liver biopsy. Surgical procedure was tailored according to the pathology found.*

Results — *A total of 18 patients were managed. The number of male and female patients was nine each. The ages of the patients ranged from one months to nine years. Most common pathology was biliary atresia (n 7). Eleven patients presented with persistent jaundice with age range from one month to 18 months. In all them HIDA scan showed failure of visualization of excretion of contrast into duodenum. All underwent laparoscopy and in seven of these biliary atresia found. There were four patients in whom extrahepatic biliary tree was found patent on cholangiogram. They were in older age range (from three months to 18 months). All had grossly cirrhotic liver.*

Three patients had choledochal cyst (two males and one female). The age ranged from 1 ½ month to 9 year. A female of 2 ¼ year presented with history of fever and failure to thrive. Ultrasound showed presence of gall stones. Cholecystectomy was done in this child. Three patients had common bile duct (CBD) pathology. All had history of fever, recurrent jaundice with dilated CBD on ultrasound. All had CBD stone / sludge. In two choledochotomy and removal of stone / sludge was done while other had cholecystectomy in addition because of inflamed gall bladder. No T tube was placed. One patient with idiopathic variety of gall stone had cholecystectomy.

Conclusions — *Ultrasound in expert hands is a good screening test in patients suspected of having surgical biliary tree pathology. HIDA scan can be safely omitted from list of investigations and laparoscopy is recommended in all cases of prolonged neonatal jaundice where surgical pathology is suspected.*

Key words — *Neonatal jaundice, Biliary atresia, Choledochal cyst.*

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INTRODUCTION:

The surgical biliary tree anomalies in paediatric patients are infrequently seen. [1] The aetiology varies with the age of the patient. In neonatal period it usually presents with persistent jaundice and infrequently with mass

while in older children there may recurrent upper abdominal pain, fever and mass with jaundice. The importance of recognizing these anomalies is particularly important in neonatal period as biliary atresia, one of the common anomalies, if recognized early and operated has the potential of sparing further liver damage. There are many overlapping features between neonatal hepatitis and biliary atresia thus distinction becomes more important because neonatal hepatitis is treated medically.

Choledochal cyst is another structural anomaly the geographical distribution of which varies.[2] It is an observation that infantile choledochal cyst behaves differently from that noted in older children.[3] The incidence of gall stones in the first decade of life is also very rare and its aetiology varies. Acute cholecystitis as a cause of acute abdomen is also rare though perforation of gall bladder is reported with this condition in paediatric population. At times some lesions present in an unusual age group with typical or atypical symptoms, awareness thus is required about their existence in a paediatric age group.

The biliary tree anomalies are thus frequently missed as they are not thought of in a workup of a patient. Even when they are kept as a cause it is not clear as to how to investigate them especially in cases of persistent jaundice. A variety of protocols are defined but there is a need of a more practical and efficient approach so that one could identify the cause and plan early treatment. The purpose of this study was to document various types of surgical biliary tree pathologies, their modes of presentations, pitfall in diagnosis and management.

METHODOLOGY:

A descriptive study was conducted in Surgical Unit B of Department of Paediatric Surgey at National Institute of Child Health Karachi from January to December 2007. It included all patients up to 12 years of age with symptoms related to biliary tree like jaundice, mass in right upper abdomen, colic etc that later on investigations found related to biliary tree.

A detailed history was taken including presence of jaundice, color of stool, mass and pain in right upper abdomen etc. All wee subjected to ultrasound as a screening test. Laboratory investigations included blood complete picture, liver function tests including bleeding profile, TORCH titre where appropriate, HIDA scan and CT scan as indicated. Laparoscopy was done in cases with persistent jaundice for operative cholangiogram and liver biopsy. Surgical procedure was tailored according to the pathology found.

RESULTS:

There were a total of 18 patients managed during the study period. There were nine males and equal number of females. Age ranged from one months to nine years. Types of pathologies found are given in table 1. Most common being biliary atresia.

There were eleven patients who presented with persistent jaundice. Their age ranged from one month to 18 months including five males and six females. All had HIDA scan done which showed failure of visualization of tracer into duodenum. All underwent laparoscopy. In seven of these patients biliary atresia was found. Three were males and four females. The age ranged from one month to three months. All had grossly cirrhotic liver. There were four patients in whom extrahepatic biliary tree was found patent on cholangiogram done at laparoscopy. Two were males and two females. They were in older age range (from three months to 18 months). All had grossly cirrhotic liver.

There were three patients with choledochal cyst. Two were males and one female. The age ranged from 1½ month to 9 year. All had ultrasound and CT scan confirmation. Hepatico-enterostomy was done in all of them. A female of 2 ¼ year presented with history of fever and failure to thrive. Ultrasound showed presence of gall stones. Cholecystectomy was done and child recovered well.

A total of three patients (2 males and one female) had common bile duct (CBD) pathology. All had history of fever and recurrent jaundice. All had dilated CBD on ultrasound. These patients had CBD stone / sludge. In two choledochotomy and removal of stone / sludge was done while the third patients underwent cholecystectomy in addition because of inflamed gall bladder. No T tube was placed. There was one patient with gallbladder stone in whom cholecystectomy was performed. There was no hematological or infective aetiology found on investigations in this patient.

Table I: Types of Pathologies

Type of Lesion	Number
Biliary Atresia	07
Patent Extrahepatic Biliary Tree (? Neonatal hepatitis)	04
Choledochal Cyst	03
CBD Stone / Sludge	03
Gall Stones	01
Total	18

DISCUSSION:

Congenital anomalies of the extrahepatic biliary tree are rare. Various types have been reported either alone or in combinations in the medical literature. Biliary atresia remains one of the commonest surgical pathologies in infancy. It is also found in this study though the number was too small. There is great overlap between this condition and neonatal hepatitis, therefore a high index of suspicion must be exercised in identifying this condition. In clinical examination color of the stool is an important observation. Normal colored stool conveniently excludes biliary atresia thus unnecessary concern as to surgical jaundice is removed. The duodenal aspiration done four hourly for presence of bile is another clinical approach to document patency of extrahepatic biliary tree.[4] Though cumbersome, but if bile stained aspirate is obtained by passing nasogastric tube into duodenum under fluoroscopic control, patient can be spared of battery of investigations. This test may be negative in patients with severe neonatal hepatitis so it can not be reliable in all cases but presence of bile excludes biliary atresia.

Ultrasound is a modality which in expert hands can clinch a diagnosis in fairly large number of cases. A classical triangular cord sign at porta hepatis is pathognomic of biliary atresia.[5] It was not reported in any of the cases in this series. A good ultrasound done in fasting state can document presence of gall bladder. If such is the case emptying of gall bladder at feeding points out to functioning and patent biliary tree. Documentation of size of gall bladder and common bile duct with a photograph must be acquired in every case. In this series gall bladder was not found on ultrasound in all cases of biliary atresia.

TORCH titre is not specific as a positive test does not exclude biliary atresia. In two cases of biliary atresia the titer was positive in our series. Extrahepatic biliary atresia (EHBA) is the most common cause of pathologic jaundice in infants. Many histological criteria are proposed for the diagnosis but still it is a challenge for the pathologist to distinguish between surgical and non surgical jaundice. Many histologic features can overlap with other causes of cholestatic hepatic diseases. Ductual proliferation and portal fibrosis are important histological features to look for. Many a times interpretation of a biopsy can be difficult and needs an experienced pathologist.[6,7] In present series this test was not used to evaluate the cause of the jaundice.

Hepatobiliary scintigraphy has poor specificity in cases of persistent jaundice. It may not differentiate between surgical and medical causes of cholestatis. A false positive rate of up to 25% to 30% have been reported.[8] Various techniques have been used to improve the postive yield of the test. By using phenobarbitone to induce liver enzyme 2 – 3 days before the procedure the false positive rate can be decreased. In a study of Spivak et al infants with low birth weight (less than 2200 g), premature infants, and those on total parenteral nutrition may not excrete the tracer into gut.[9] Same is the case with infants having severe hepatitis. In four patients of our study HIDA scan was negative though on cholangiogram biliary tree was found patent.

Laparoscopy is an en excellent modality both for diagnostic and therapeutic purposes.[8] Aspiration of visibly dilated gall bladder and per operative cholangiogram facilitates the diagnosis. Cholangiogram done by cannulating gallbladder must demonstrate the entire biliary tree. A laparoscopic assisted single port technique is described by Jan et al for performing cholangiogram. [10] An attempt must be made to delineate the proximal intrahepatic tree by application of a distal vascular clamp. Liver morphology can also be gauged and biopsy taken. Where facilities and skills are present portoenterostomy (Kasai' s procedure) can be performed in cases of biliary atresia.

CT scan is routinely used in hepatobiliary conditions and is cost effective as well. Newer modalities such as ERCP and MRCP have been used at times for pathologies like choledochal cyst and idiopathic CBD dilatation. They provide excellent view of the anomaly and treatment can be planned accordingly. We used CT scan in patients with choledochal cysts and those in whom ultrasound showed dilated CBD. In all these cases treatment got facilitated. The dilated bile duct is a term not agreed upon by all. [11] Various dimensions have been proposed to define upper limit of CBD in relation to age groups. Hernanz-Schulman et al reported that the diameter of the CBD should be less than 1.6 mm in infancy and less than 3 mm in childhood on ultrasound.[12] Siegel reported that on ultrasound a visible intrahepatic biliary duct and a CBD diameter more than 2 mm in infancy, greater than 4 mm in childhood, and greater than 7 mm after adolescence are abnormal.[13] In three of our patients grossly dilated CBD was visible on CT scan. They all underwent CBD exploration and sludge and stones were removed. No T- tube was placed following choledochotomy and children recovered well. Use of T – tube is also a controversial area but there are reports even from adult series where T – tube placement is not done routinely after CBD exploration.[14]

In one study Lee et al after reviewing ultrasound of 162 children with pre defined inclusion criteria found that all dilatations of the bile ducts with intrahepatic involvement are anomalous biliary tree disease and surgery is indicated.[15] Dilatations of the extrahepatic

biliary tree (fusiform type in particular), are not necessarily diagnostic of choledochal cyst. Of the extrahepatic biliary duct dilatations in their study, all choledochal cysts were greater than 10 mm in diameter. Patients with CBD dilatation of less than 7 mm in diameter according to them be investigated and followed before surgery is undertaken. The term forme fruste choledochal cyst (FFCC) was first used by Lily et al and similar pathology has been described by Okada et al as common channel syndrome.[16,17] This condition is now considered to be a variant of choledochal cyst. The maximum normal length of the common pancreaticobiliary channel in infants has been reported as 3 to 4 mm; it increases with age to a maximum of 5 mm in adolescents as cited by Kumar et al.[18,19] In our patients there was no distal obstruction and simple irrigation of CBD was enough to treat the symptoms. Patients remained symptom free in follow up as well. Patients with choledochal cyst and gall stones presented with typical symptoms and underwent standard surgical protocol. The recovery in all of them was uneventful even in an infant with choledochal cyst which is considered a different pathology in comparison with that presenting in older children.

CONCLUSIONS:

Overall occurrence of biliary lesions remained low in paediatric age group. History and clinical examination have high specificity. An experienced ultrasonologist can help in documenting patent extrahepatic biliary tree. HIDA is not a sensitive investigation as differentiation between biliary atresia and neonatal hepatitis can not be made in every case. Laparoscopy helps in diagnosis and in experienced hands definitive procedure can be performed in the same sitting.

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