Choledochal cysts in children: Current issues and experience at a Pediatric surgical unit in Baghdad

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Abstract

Background: C
holedochal cysts are somewhat rare congenital anomalies of the biliary system, but proper, timely diagnosis and treatment prevents grave complications.

Aim
: to evaluate cases diagnosed at a major pediatric surgical center in Baghdad, over a one year period.

Methods
: During a one year period, eleven pediatric cases were diagnosed with different forms of choledochal cysts, their clinical presentations, imaging work up were reviewed according to their ages. In addition to their treatment and outcomes. **Results:** Females formed 81.8% of the diagnosed cases. About two thirds of the cases were diagnosed below 1 year of age, mainly with cholestasis, the remaining older group patients, presented mainly with abdominal pain. Ultrasound examination was diagnostic in all but one, in whom the diagnosis was fulfilled by CT scan
.Total cyst excision, was done in 8 cases, one with additional Kassai surgery. No postoperative complications, except one child who was treated by internal drainage procedure. No malignancies documented

Conclusion:
Choledochal cyst is not a very rare disease in Iraqi pediatric population. The general demographic data follows the international records. Efforts are needed to enhance prenatal diagnosis of this anomaly, and better follow up of diagnosed cases.

Introduction: Choledochal cysts are congenital anomalies of the biliary tract characterized by cystic dilatation of the extrahepatic biliary tree, intrahepatic biliary radicals or both. The first anatomic study of a choledochal cyst in the Western literature was published Vater and Ezler in 1723. Douglas is credited with the first clinical report, on a 17-year-old girl who presented with intermittent abdominal pain, jaundice, fever, and a palpable abdominal mass.[1]

Alonso-Lej et al provided the first systematic description of choledochal cysts, and classified choledochal cyst into 3 types based on the clinical and anatomic findings in 96 cases.[2] The classification system for choledochal cysts was further refined by Todani and colleagues and currently *.

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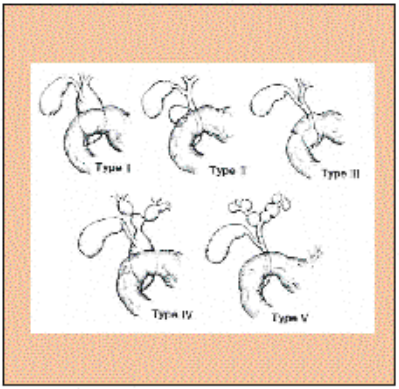
includes 5 major types (fig 1).[3] The condition is a relatively rare abnormality in western population of 1:13000-1:15000 while it is far more common in the east 1:1000 in Japan[4]. The etiology of choledochal cysts is still unknown, but they are generally considered to be congenital [5].The common long channel theory is the most widely accepted one, where the abnormal Pancreatico-biliary malunion (PBMU) leads to pancreatic reflux into the biliary ducts causing damage and subsequent dilatation of the ducts [6,7]. On the other hand there are increasing numbers of antenatally diagnosed choledochal cysts reported [8], and since the secretory response of the human pancreas to secretagogues is acquired after the perinatal period, thus, a choledochal cyst due to amylase reflux is unlikely during this period because the enzyme is absent prior to birth [9,10]

Choledochal cyst may be diagnosed at any age but two thirds of the patients are diagnosed before ten years of age [11]. Choledochal cyst was reported at 78 years of age[12]

.Patients with choledochal cyst diagnosed before I year of age should undergo surgery as soon as possible to avoid complications; as ruptured CC cyst , liver cirrhosis which was reported in a newborn as early as 10 days old [13], and liver failure due to untreated choledochal cyst in a 5 mo old infant [14]



Methods: Over a one year period, from October2008 -Oct 2009, eleven patients were treated for different forms of choledochal cysts (9 females and 2 males), at the Children welfare teaching hospital /The Medical City in Baghdad. It was a prospective analysis of the clinical presentation of the disease, diagnostic measures, treatment and complications.

The ages of the patients ranged from 4wks 9 years.According to Todani classification (diagram I) [1]

, eight cases were type I (one case with combined CC and atresia of the extrahepatic ducts), one case was type IVa (intrahepatic & extrahepatic cystic dilatation with features suggestive of hepatic fibrosis) and one case with Caroli syndrome (type V).

Total excision of the cyst done up to level of hepatic ductal bifurcation and to the most distal caudal extension of the cyst ,then anastomosis of jeunal loop to CBD at level of transection with infracolic enteroenterostomy was performed .

Results:

For the purpose of comparing the clinical manifestations, patients were divided into GroupI: under 1 year of age; seven cases 7/11 (63.6%), included was a 2yr old child who was operated on at 4 mo of age for choledochal cyst, presented now at 2yr age with intestinal obstruction. There were 2 male babies in this group only, the rest were females.

GroupII: over 1 year of age; (four cases 4/11), all were females.

Females formed 81.8% of the whole diagnosed cases.

Clinical manifestations:Jaundice was a major presenting feature in group I patients. It was present in 6/7 (85.7%) of them, fever was present in 5/7 (71.4%) of the patients (as a feature of cholangitis), clay colored stool was present in one case, abdominal mass and multiple biliary stones were noted in one 4 mo old patient.

While abdominal pain was a major presenting feature in group II patients. It was present in 3/4 (75%) of them (a six year old girl presented with recurrent pancreatitis, another 5 year old girl showed the classical triad of jaundice, abdominal pain and mass), fever was present in 3/4 (75%) of them, while jaundice was seen in only 1/4 (33.3%) of group II patients. One patient had features of portal hypertension and liver fibrosis (the one with Caroli syndrome). Family history was negative for similar conditions, and no other congenital anomalies were documented in all studied patients.
The clinical characteristics of each group are listed in table 1 and 2.

Imaging procedures:
Ultrasound examination was used in all patients and it was successfully diagnostic, except a 6 year old girl where a pancreatic cyst was suspected on U/S, and the diagnosis of CC was documented on MRI. MRI was applied in a second case (the one with type IV choledochal cyst), CT scan was done to two patients, and ERCP was done to one patient (5 year old) with recurrent pancreatitis.

Surgical treatment & complications:
Seven patients were treated by total excision of the cyst and hepaticojejunostomy, while the 2 yr old child who lately presented with intestinal obstruction, was treated with a sort of internal drainage procedures, and another 3 month old baby girl was treated with both cystectomy and Kassai procedure. There were no operative deaths. No post operative complications during the period of follow up, that ranged from 2 - 8 mos, except the child with intestinal obstruction that presented 16 mo after the initial surgery, adhesive bands were found on laparotomy. The patients with type IV and V disease were kept on conservative measures only.

No evidence of malignancy was detected.

Table 1: characteristics of group I patients

Group I	age	sex	presentation	complications
1	1 mo	female	Jaundice + fever	
2	6 wk	male	Jaundice + fever	
3	6 wk	female	Jaundice+ mass+ fever	
4	4 mo	male	Ascites+ peritonitis	Late postop. adhesions
5	4 mo	female	Jaundice + mass+ fever+ cystic & ductal stones	
6	5 mo	female	Jaundice	
7	3 mo	female	Jaundice	Associated extrahepatic biliary atresia, persistent jaundice postoperatively, but with colored stool

Table 2: Characteristucs of group II patients

Group II	Age	Sex	Presentation	complications
1	4 yr	female	Abdominal pain , ascites, fever (peritonitis)	Portal hypertension
2	6 yr	female	Recurrent pancreatitis	
3	5 yr	female	Pain, jaundice, mass, fever	
4	20 mo	female	Cholangitis	

Discussion:

Congenital cystic dilatation of the common bile duct (choledochal cyst) is a rare disease in the Western countries, and most of the case reports came from Asia, while two thirds of the reported cases came from Japan[6]. The international incidence of the disease is about 1 in 2000000, but it is about four times more frequent in the yellow race [6]

.The Children's welfare hospital pediatric surgical unit, is one of the main units serving pediatric surgical problems in Baghdad and surrounding governorates, another surgical unit in the hospital of Gastrointestinal and liver diseases and that of the Medical City (Baghdad Teaching hospital), although the last two deal mainly with adult problems . There are probably two more centers dealing with pediatric surgical problems in Iraq, one in the North, and possibly another in the south of the country.The eleven studied cases were collected within a one year period. All the children, came from Baghdad or nearby surrounding governorates, so the general incidence of this anomaly maybe at least double this figure allover Iraq, with an overall birth rate in Iraq that is estimated to be 1.2 million births per year, Iraq seems to be of the higher incidence countries. A Jordanian report in 2006 [7], thought that probably Jordan had a lower incidence of CCD in comparison with Asia, where only9 cases were treated at the Center of the Royal Medical services, over a 24 yr period! but possibly it was the smaller population size and/or diagnostic and referral problems that yielded this result. They even had not demonstrated any sex difference in their reported cases, while it is reported that the female to male ratio is 4:1[8,9]

. We had a similar ratio with female preponderance in our studied cases.Two thirds of the studied cases were diagnosed during the first year of life. Antenatal diagnosis is now feasible in many places [10,11], and that permits early surgical treatment, before recurrent cholangitis and pericystic inflammation occur, and makes cyst excision easier[12]

. Unfortunately none of our patients was diagnosed antenatally.Younger patients, under 1 year old (group I) presented mainly with obstructive jaundice, while older children (group II) mainly presented with abdominal pain, suggesting biliary pancreatitis, a feature repeatedly demonstrated in reports of choledochal anomalies

[17,21]In both groups fever was a common presentation (due to cholangitis and /or pancreatitis), it affected 70% of the cases, which is much higher than what is noted in most medical reports, possibly due to delay in the diagnosis in our patients, allowing them a longer period of complications. The classical

triad of abdominal mass, abdominal pain and jaundice, which is the known presentation of CC is reported in 25-30% of cases [3]
, it was also seen in 3/11 of our patients.Choledochal cyst should be always be considered in the differential diagnosis of jaundice and pancreatitis. The golden diagnostic tools for CC are ultrasound and CT scan. MRI and MRCP were also reported to be very helpful to define the anatomy of the biliary system [15], but it may not be a sensitive tool in pediatric cases as it is in adults, where ultrasound has a preeminent role [16]. Common bile duct measures less than 3.5 mm in healthy children and less than 2mm in infants [17]. This will help to detect dilatations of the common bile duct through various imaging techniques. Differentiating neonatal CC from correctable biliary atresia is difficult; because the distal end of the neonatal CC is often totally obstructed, an ultrasound can be helpful in demonstrating the CC being larger, intrahepatic ducts usually dilated and gall bladders are not atretic in patients with CC [18]
.Ultrasound examination was sufficiently diagnostic in most of our patients, but unluckily prenatal diagnosis was not practiced.
In the past many surgical procedures were described for the treatment of choledochal cyst like (external drainage, cyst marsupialization, sphincteroplasty and choledocho-cysto-deudenostomy). Some of these procedures failed to treat the patients and others were associated with high morbidity and mortality due to multiple postoperative complications, like strictures, recurrent cholangitis, and malignant transformation, as reported in many studies from all over the world [19]. Total excision of the choledochal cyst with biliary reconstruction has now been a widely accepted procedure even in infants and children [10,11]. Cyst excision eliminates a reservoir for bile stasis and biliary obstruction; reduce the incidence of cholangitis, stone formation and future malignancy [11]. We did receive one of the children who were previously treated with internal drainage procedure without cyst excision presented 16 months later with intestinal obstruction due to adhesive bands while the rest of the patients did not present with signific
ant postoperative complications.Chokshi et al reported on laproscopic management of CCs in children and found that laproscopic resection of CC and Roux-en-y hepaticojejunostomy is an excellent treatment option, after reviewing 9 patients in the pediatric age group with this technique, in the period 2003-2007 with no intraoperative complications and very good recovery rate
"The patient with type IV choledochal cyst was treated conservatively and was thought to be inoperable; however Kawarda has treated 13 patients with this type of defect by resection of extrahepatic bile duct including pancreatobiliary maljunction and hepaticojejunostomy, because it is frequently associated with malignancy and to improve long term survival"

Conclusions and recommendations:

1- Choledochal cyst malformation is not a rare disease in Iraqi infants and children, possibly Iraq are one of the high incidence areas
.2- Cholestasis in infancy, and recurrent abdominal pain in the older child should lead to the suspicion of biliary tract malformation, maybe due to different pathogeneses in these two groups
.3- Cyst excision and biliary reconstruction is the recommended procedure with the least morbidity and mortality.
4- Efforts should be enhanced to orient physicians (sonographists) for antenatal diagnosis of biliary tract malformations.

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