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Case Series

Surgery Section

External Drainage of Giant Infantile Choledochal Cyst before Definitive Repair: Is it Worth?

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ABSTRACT

Infantile Choledochal Cysts (IFCC) usually present with jaundice, acholic stool and abdominal lump or abdominal distension. If the surgical intervention is delayed, they rapidly progress to liver fibrosis which is considered to be irreversible if progressed to cirrhosis. We present the data of four cases (aged one month to seven months) of IFCC presented with cholangitis managed in one surgical unit in last two years. In one case, cholangitis was treated with prolonged antibiotic course before definitive repair whereas in rest, external drainage of cyst was done in addition to intravenous antibiotic to treat cholangitis. All the infants had features of cholangitis at time of presentation. Total leucocyte count ranged from 18x1000/UL to 30.6x1000/UL. Total bilirubin level at presentation ranged from 8.2 mg/dl to 18 mg/dl and Prothrombin time (INR) ranged from 1.33 to 1.9. Hepatic fibrosis was observed in all cases but cirrhosis was observed in only one case. There was no mortality but one patient had postoperative complication with prolonged hospital stay. External drainage helps in early recovery from cholangitis and better optimization of liver function. It also delays further progression to liver fibrosis by relieving the biliary outflow obstruction while waiting for definitive repair.

Keywords: Antibacterial agents, Bilirubin, Liver cirrhosis

IFCC differs from cyst in children (classical children) and adults in presentation, pathology, complication and outcome. Todani T et al., have characterized the IFCC as follows: cystic choledochal dilatation, abdominal mass with jaundice and acholic stools, no symptomatic association with acute pancreatitis and a low amylase level in bile [1]. We are presenting four cases of huge IFCC presented in newborns and infants of which three were managed with external biliary drainage before definitive treatment.

CASE 1

A seven-month-old female child presented with high grade fever, jaundice, passing pale stool, abdominal distension and vomiting since five days. Clinical examination revealed that she was malnourished, had abdominal distension with palpable liver and free fluid. History of jaundice and passing pale stool was noticed since age of one month. Haematological, biochemical and liver function tests were abnormal [Table/Fig-1]. Ultrasound and Magenetic Resonance Cholangiopancreatography (MRCP) showed Todani Type–I Choledochal Cyst (CC) [Table/Fig-2]. Patient was initially managed with intravenous fluid, antibiotics and blood products and considered for excision of the cyst and Rous – en – Y hepaticojejunostomy (RYHJ) after stabilization (after two weeks).

On exploration, it was a large cyst with grossly distended gall bladder and cystic duct. Cyst was oedematous and inflamed with lot of free fluid.

Dissection of the cyst was difficult because of inflammation and tissue oedema; hence surgery was prolonged (more than six hours). Liver was hard and dark, Histopathology Examination (HPE) of liver showed distorted hepatic architecture with bridging fibrosis, ductular proliferation, widening of portal tracts and formation of nodule. Postoperative course was complicated, needed ventilator support for more than 24 hours, developed wound infection and partial wound dehiscence, chest infection, fever, hypoalbunemia and electrolyte imbalance due to high drain output. Patient was allowed oral feed on eight Postoperative Day (POD) and was discharged on 20th POD due to issue related to high drain output and sepsis. Liver function was on improving trend and almost normalized in follow up after one year [Table/Fig-3]. Patient developed incisional hernia but now thriving well and waiting for repair.

CASE 2

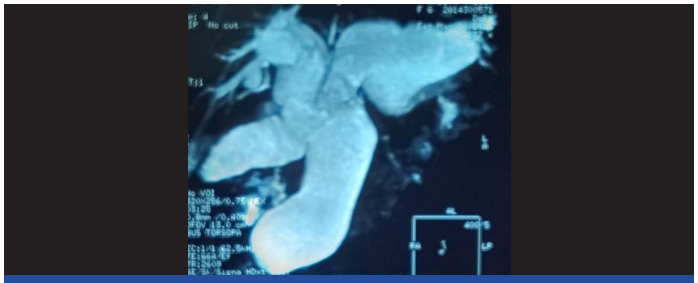
One-month-old male child weighing 3 kg, presented with fever, respiratory distress, jaundice, passing pale stool, abdominal distension [Table/Fig-4] and failure to thrive. Haematological and biochemical investigation are tabulated in [Table/Fig-1]. Ultrasound abdomen showed large Todani Type-I, CC with coarse echo texture of liver with minimal free fluid in the peritoneal cavity. MRCP showed huge CC with minimal Intrahepatic Biliary Radicle (IHBR) [Table/Fig-5]. Considering the general condition, deranged Liver Function Test (LFT), very large size of the cyst, complete excision of the cyst with bilioenteric anastomosis did not seem safe even after a week on intravenous antibiotics and other supportive medication. The patient was planned for open cholecystostomy as percutaneous trans-hepatic biliary drainage was difficult because hepatic ducts were not significantly dilated. Endoscopic Nasobiliary Drainage (ENBD) was not possible at our center in this age group hence cholecystostomy was performed via very small subcostal incision (around 2 cm). A 6 fr. Foley catheter was negotiated into the cyst [Table/Fig-6], liver was hard, looked dark and green and intraperitoneal contents were stained yellow [Table/Fig-6]. Cholecystostomy drained 400 ml bile at the time of operation and drainage via cholecystostomy was ranged between 260 to 300 ml per day which was replaced meticulously by appropriate fluid. LFT was normalized after three weeks of drainage [Table/Fig-3] and patient was taken for definitive surgery. Excision of the cyst was easy because it was deflated and not inflamed. Complete excision of the cyst with RYHJ was done. HPE of liver showed ductular and parenchymal cholestasis, mild ductular proliferation no evidence of cirrhosis. Oral feed was allowed on 5th POD and discharged on eight POD. Patient is thriving well on follow up.

CASE 3

A five-month-old female child presented with abdominal distension, vomiting, jaundice, passing clay color stool and failure to thrive. Examination revealed palpable liver with lump in right hypochondrium with free fluid and weighing 5.5 kg. Haematological and biochemical parameter were deranged [Table/Fig-1] LFT and coagulation was deranged. Ultrasound and MRCP showed Todani Type –I CC. Since patient had cholangitis, Percutaneous Transhepatic Billary Drainage

Parameters	Case – 1	Case – 2	Case – 3	Case – 4
Age	Seven months	One month	Five months	One month
Sex	Female	Male	Female	Male
Weight	5 kg	3 kg	5.5 kg	3 kg
Presenting feature	Lump, fever, jaundice, acholic stool, excessive cry and vomiting	Lump, jaundice, acholic stool, abdominal distension, respiratory distress	Lump, fever, acholic stool, fever	Lump, jaundice, acholic stool, abdominal distension
Hematlogical investigation at presentation	Hb- 10.9 gm% TLC- 30.6 x1000/ul PLT-158 x1000/cmm	Hb- 8.7 gm% TLC-28.8 x1000/ul PLT-281 x1000/cmm	Hb- 8.7 gm% TLC-18.8 x1000/ul PLT-231 x1000/cmm	Hb- 13.9 gm% TLC- 23.1 x1000/ul PLT - 250 x1000/cmm
Type of cyst	one	Ia (very large cyst with volume of cyst at presentation on ultrasound is a round 350 ml)	Ia	Ia (very large cyst with volume of cyst around 400 ml)
Complications at presentation	Cholangitis, features of portal hypertension at time of exploration and coagulation deranged	Cholangitis very high bilirubin level, coagulation deranged	Cholangitis with deranged coagulation	Very high bilirubin level with coagulation deranged
Procedure	Exicision of cyst with RYHJ	Cholecystostomy followed by RYHJ after one month	PTBD followed by RYHJ after one month	Cholecystostomy waiting for surgery
Cystic fluid amylase and serum amylase on day of surgery	254 u/l, 56 u/l	180 u/l, 45 u/l	92 u/l, 75 u/l	86 u/l, 45 u/l
Intraoperative time during definitive surgery	Eight hours with significant blood loss	Five hours	Five hours	Not operated
Postoperative complication	Chest infection, ascites, wound infection, wound dehiscence, long hospital stay (three weeks)	Minor wound infection (hospital stay 10 days)	None, hospital stay seven days	Not operated

[Table/Fig-1]: Showing presenting feature, liver function test, hematological test, ultrasound finding, MRCP finding at the time of presentation.



[Table/Fig-2]: Showing dilated left and right hepatic duct with dilated CBD and distended gall bladder.

(PTBD) was done in this case for biliary drainage because hepatic duct was prominent. Excision of the cyst with RYHJ was done after three weeks of drainage when the liver function was improved [Table/Fig-3].

At time of definitive repair, liver was firm and coarse in echotexture. Intraoperative liver biopsy showed ductular proliferation with no evidence of cirrhosis. Postoperative course was uneventful and patient was discharged on 10th POD.

CASE 4

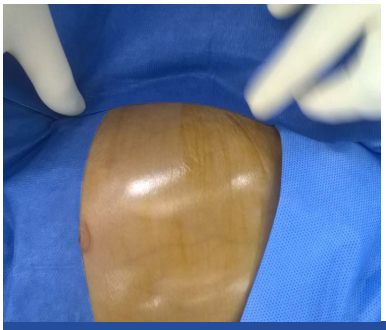
One-month-old child presented with jaundice, passing yellow color stool and abdominal lump. Investigation revealed jaundice, elevated transaminase, hypoaluminemia and coagulation abnormality. Ultrasound and MRCP showed Todani Type Ia CC with free fluid [Table/Fig-7]. Patient had respiratory distress because of large lump and hence, was considered for cholecsytostomy after stabilization. Cholecsytostomy drained 400 ml of bile at time of insertion and thereafter draining 150 to 200 ml per day. Patient was discharged on 5th POD and on first follow up after three weeks liver function was imporving [Table/Fig-3]. Liver biopsy at time of the cholecystostomy showed broadening of the portal tract and mild ductular proliferation with no evidence of cirrhosis.

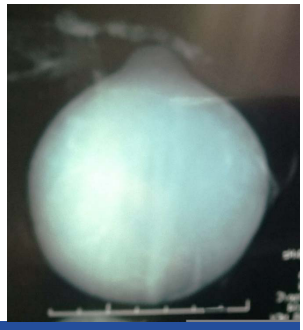
DISCUSSION

CC in newborns and infants is different when compared to the disease in children and adults and seems to have clinical and pathological features similar to correctable biliary atresia [2]. The key issue in IFCC is to differentiate it with Correctable Biliary Atresia (CBA) [3] and appropriate timing for intervention.

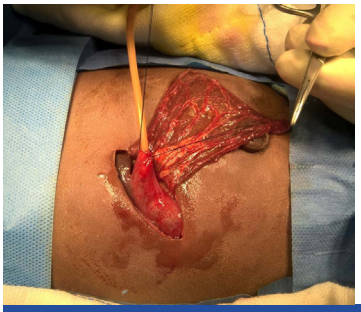
Case	Liver function test	At presentation	At surgery/ or first follow up after drainage	Follow up one month	Follow up three months
Case-1	T. Bil	12.2	5.2	1.2	1
	Direct Bil	5.6	3.3	0.6	0.6
	SGPT	90	50	29	27
	SGOT	65	105	49	41
	ALP	381	335	285	163
	GGT	111	205	82	29
	S. Albumin	3	3.4	2.9	3.6
Case -2	T. Bil	14.6	2.1	1	0.57
	Direct Bil	10.4	1.5	0.5	-
	SGOT	92	196	61	61
	SGPT	83	209	46	44
	ALP	404	365	150	147
	GGT	367	267	289	190
	S. Albumin	2.5	3.9	3.9	4
Case – 3	T. Bil	8.2	0.8	0.7	0.6
	Direct Bil	5.3		0.3	0.3
	SGOT	239	119	84	41
	SGPT	129	54	54	28
	ALP	642	586	559	295
	GGT	609	460	146	16
	S.Albumin	3.2	4.1	4.1	4.3
Case – 4 *	T. Bil	18	3.8		
	Direct Bil	14	2.6		
	SGOT	675	556		
	SGPT	1332	417		
	ALP	497	407		
	GGT	346	256		
	S. Albumin	3.3	3.6		

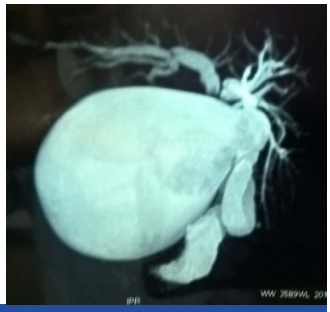
[Table/Fig-3]: Trends of liver function test at time of admission, surgery and follow up.
ALP: Alkaline phosphatase
GGT; Gamma –glutamyltranspeptidase
S. Albumin: Serum Albumin
T. Bil : Total Bilirubin
Direct Bil: Congugated Bilirubin, *Definitive repair not done till date





[Table/Fig-4]: Showing abdominal distension due to giant choledochal cyst. **[Table/Fig-5]:** MRCP showing large choledochal cyst with minimal dilatation of the right and left hepatic duct.





[Table/Fig-6]: Intraoperative picture showing normal sized gall bladder with yellowish discoloration of the omentum and liver. **[Table/Fig-7]:** MRCP showing giant choledochal cyst with grossly dilated gall bladder.

IFCC can be differentiated preoperatively from CBA by ultrasound [4]. The cysts in case of IFCC are: larger and thick walled in comparison to the CBA. Intrahepatic bile ducts in cystic biliary atresia Billary Cystic Atresia (BCA) are hypoplastic and irregular whereas; those in CC are well formed and often dilated, if the size of the cyst is small (< 8 mm) it favours biliary atresia and bigger size favours IFCC. Gall bladder is usually atretic or small in the cases of Billary Cystic Malformation (BCM) whereas, it is almost normal in cases of IFCC. The second issue is appropriate timing for the surgery which is important because of higher incidence of liver fibrosis or cirrhosis in cases of IFCC in comparison to CC in children or adult at time of presentation and it was that liver fibrosis is more common and severe in infants and children at the time of presentation [4].

Aggerwal N et al., has observed 71% of infants had either evidence of liver fibrosis or cirrhosis in comparison to only 30% those presented after one year in their study [5]. Around 2%-10% cases of liver fibrosis due to obstructive jaundice in cases of CC develop cirrhosis [2,6,7], since liver cirrhosis is considered as irreversible disorder, early definitive surgery is generally advocated.

There is enough literature to support that the clinical manifestation, pathology and aetiopathogenesis of IFCC differ from the CC in children and adults [2].

Aetiopathology of IFCC cannot be explained by hypothesis of Babbitt DP, which is based on the pathophysiological consequence of reflux of activated proteolytic pancreatic enzymes on the biliary tract wall [8]. But can be explained by the hypothesis that obstruction of bile duct leads to increased proximal bile duct pressure [9] and eventual dilatation, initially of the extrahepatic segment and subsequently the intrahepatic component. Infantile CC presents with jaundice, acholic stool and lump whereas, children presents with recurrent abdominal pain, vomiting, jaundice and cholangitis. Cystic fluid amylase level were normal in our series and was similar to the findings of previously reported series of IFCC whereas in patients presenting beyond infancy, it was significantly raised [2,5,9]. All these findings support the fact that IFCC is clinically and pathologically different form CC in children and adults. If CC in children is left untreated, it can cause morbidity and mortality from recurrent cholangitis, pancreatitis, sepsis, liver abscesses, and cholangiocarcinoma [10] whereas in cases of newborns and infants, it will either rapidly progress to cirrhosis due to obstructive jaundice [6,7,10] or result in rupture of the cyst because of high intracystic pressure owing to blind distal end [10]. Most of IFCC are either Type-Ia or less commonly Type–IV according to Todani's classification, as was seen in our series (three were Type Ia and one was Type-IV).

Objectives of surgical treatment are total excision of the cyst with patent bilioenteric drainage in form of either hepatico-duodenostomy or RYHJ. We favour RYHJ and all the patients are doing well in follow up with no complication related to hepatobiliary system in long term follow up. In present series, in three cases, cyst was very large (maximum transverse diameter was more than 8 cm and volume was more than 400 ml), and all had signs of cholangitis at presentation.

In first case, we treated the cholangitis with intravenous antibiotics, (three weeks) and coagulation was normalized with vitamin K and blood products. Definitive repair was done after three weeks when the features of cholangitis subsided and hematological parameter and coagulation was normalized. Postoperative course was very complicated as evident by case history. In view of the complication observed in first case, we decided to drain the cyst externally to treat cholangitis (in two cases, we did open cholecystostomy and in one case PTBD was done to drain the cyst) in order to treat cholangitis and properly optimize the liver function, [Table/Fig-3] prior to definitive repair. We presume that, the delay of three weeks during the optimization prior to surgery in first case, further damaged the sick liver, and this delay might have caused the progression to liver fibrosis, though we have no definitive evidence because liver biopsy was not done at the time of presentation. [Table/Fig-3] shows that liver function was almost normalized within three to four weeks of the drainage procedure in three cases whereas in case one, it was not optimized at the time of definitive repair, though, it was normalized in all cases at follow up of one year. Intraoperative and postoperative course was uneventful in cases that underwent drainage procedure before definitive surgery and were discharged within 10 days of surgery. Many authors had advocated for early definitive repair of the cyst presenting in neonatal period or in infancy and consider it safe [2,6,7,10]. We did not find details of intraoperative technical difficulties, and problem faced by the treating team in postoperative period after definitive surgery, especially for the giant choledochal cyst as in our series except the case reported by Tetsuya where a five-month-old child, developed; wound disruption requiring reoperation, sepsis, disseminated intravascular coagulation, after definitive operation for giant choledochal cyst presented with complication (altered liver function and cholangitis). In present series, we faced such complication in one patient (Case 1), where liver function was not optimized at time of definitive repair, but in rest of the cases there were no intra or perioperative complication. We observe that, drainage of cyst helped in early control on cholangitis, better optimization of the liver function and coagulation, easier dissection of the cyst at time of definitive repair, and last but not the least it does not further damage the sick liver while waiting for the definitive repair by relieving the bile outflow obstruction.

CONCLUSION

Infantile choledochal cyst must be treated as soon as possible but if they presents with complications, it is better to drain the cyst to treat cholangitis instead of treating it alone with antibiotics to avoid further damage of already sick liver.

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