Giant choledochal cyst in a 17 year old female at a teaching hospital in Uganda: A Case Report and review of literature

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Abstract

Choledochal cyst is a rare congenital dilatation of the common bile duct usually symptomatic within the first year of life with rare presentation in adulthood. Herein, we present a 17-year-old female with type 1A choledochal cyst who was managed by surgical excision, hepatico-jejunostomy and jejunojejunostomy who recovered uneventfully.

Giant choledochal cyst in a 17 year old adult girl at a teaching hospital in Uganda: A Case Report and review of literature

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Abstract

Background:

Choledochal cysts are rare congenital bile duct anomalies that lead to cystic dilatations of the biliary tree involving either extra-hepatic biliary radicles, intra-hepatic radicles or both. This illness is very rare in Africa with Akinyika et al., in Nigeria reporting only 3 cases in 18 years. Surgical management of the cyst is the main form of treatment since it reduces the risk of long term complications like malignancy, cholangitis and gallstones. Type I lesion is the most common accounting for 60-90%.

Case:

A 17 year old female presented with 4 months history of progressive abdominal distension associated with abdominal pain, yellowing discoloration of eyes, and on and off constipation. Abdominal CT-scan showed a huge cystic mass in the right upper quadrant extending inferiorly to the right lumbar region. Complete cyst excision of type IA was done plus Cholecystectomy in addition to bilioenteric reconstruction.

Conclusion:

Giant choledochal cyst is a very rare disease which can be diagnosed using abdominal CT scan and abdominal ultrasound scan. Use of ERCP and MRCP techniques is increasing for the non-invasive diagnosis of choledochal cysts however they are still rare and expensive in our local setting. Complete surgical excision reduces the chances of pancreatitis and biliary malignant transformation. This is one of the first reported cases of a young adult in the literature of giant choledochal cysts to be managed successfully in any African nation.

Key words : Giant choledochal cyst, hepaticojejeunostomy, todani classification

Introduction:

Choledochal cysts are infrequent congenital malformations characterized by cystic dilatations of the biliary tree that was first described by Vater and Ezler in 1723(1). These dilatations can be extra-hepatic, intrahepatic or both and in this case, it was extra-hepatic. The incidence of choledochal cysts in Asian population is 1 in 1000 live births with predominance in females(2). In western countries, the incidence ranges from 1:100,000 to 1:150,000 in the United States, 1 in 2 million in the United Kingdom. In Africa, there is insufficient data documented about giant choledochal cysts. More than 60% present in the 1st year of life and 20% in the adulthood(3).

Initial classification by Alonso-Lej et al.in 1959 described 3 types of choledochal cysts, type I-III(4). In 1977, Todani and others modified the original Alonso-Lej classification to include type IV and V (Figure 1). Type I of choledochal cysts are further sub-classified into 3 types. Type IA, cystic dilatation of entire extrahepatic biliary tree, type IB is focal, segmental dilatation of extra-hepatic biliary tree and type IC is fusiform dilatation of entire extra-hepatic biliary tree. Type II describes discrete extra-hepatic bile duct diverticulum while type III often has bulging into duodenal lumen(choledochocele).Type IV A multiple sites of dilatation, both extra and intrahepatic biliary tree and Type IVB has multiple sites of dilatation of extra-hepatic bile duct only while type V has cystic dilatation of only intrahepatic biliary tree(Caroli disease) (5).

Choledochal cysts in children and adults behave differently with type I being common in children and type IV in adults. Anomalies of pancreatic duct and other associated hepatobiliary problems are more seen in adults than in children(6). No strong unifying etiological theory exits for choledochal cysts but some series published by Miyayo and yamataka in 1977, documented an anomalous junctions in 90-100% of patients with choledochal cysts(7). This abnormal communication allows the mixing of bile and pancreatic juices activating pancreatic pro-enzymes that later damages and weakens the bile duct wall resulting in formation of choledochal cysts (8).

The primary diagnostic modality is by abdominal ultrasound scan followed by CT scan and the MRI. The associated anomalies are biliary atresia, gallbladder atresia, hepatic fibrosis and those of pancreatico-biliary ductal system. ERCP and MRCP have a conclusive role in confirming ultrasound scan diagnosis. This is also done to evaluate anatomy and identify complications related to the cyst(5). In this case, a CT scan was done to confirm type IA giant choledochal cyst.

This choledochal cyst is usually associated with complications that range from cholecystitis, cholangitis, pancreatitis, stone formation and malignancy. Prevalence of biliary malignancy is found to be around 30% of the cases, this increase with age and is commonly higher in those with type I and type IV choledochal cysts(9).

The treatment of choledochal cyst is an initial control of complications then followed by the primary surgery of the cause. Presently the most popular and accepted surgical intervention is by total excision of the choledochal cyst and restore the biliary enteric drainage into the duodenum either via Roux-en-Y hepatico-jejunostomy or Braun hepatico-jejunostomy. The later approach was used in our patient(10).

A laparoscopic approach is also currently being explored since it offers less invasive of the surgical technique approach for the management of choledochal cyst in children (11).

We report a case of a 17 year old female who was diagnosed and managed for a giant choledochal cyst type IA at Mbarara Regional Referral Hospital, Uganda

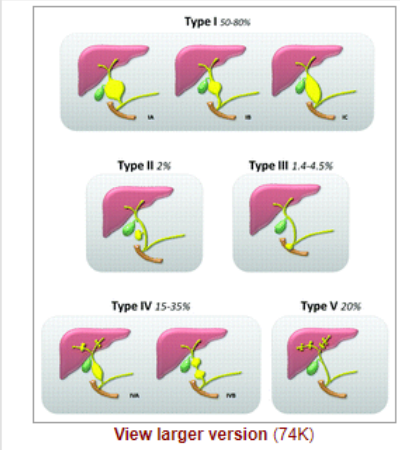


Fig. 9 ???Schematics show Todani modification of Alonso-Lej classification and relative percentage of occurrence of each type of choledochal cyst. Type IA is marked cystic dilatation of entire extrahepatic bile duct; IB, focal segmental dilatation of extrahepatic bile duct, which can occur at any level but is usually distal to cystic duct insertion; IC, smooth fusiform dilatation of entire extrahepatic bile duct; II, discrete diverticulum of extrahepatic bile duct; III, dilatation of distal common bile duct confined to wall of duodenum, often bulging into duodenal lumen (choledochocele); IVA, multiple sites of dilatation of both extrahepatic and intrahepatic biliary tree; IVB, multiple sites of dilatation of extrahepatic bile duct only (string-of-beads appearance); V, multiple sites of saccular or cystic dilatation of only intrahepatic biliary tree (Caroli disease or communicating cavernous ectasia) [17].

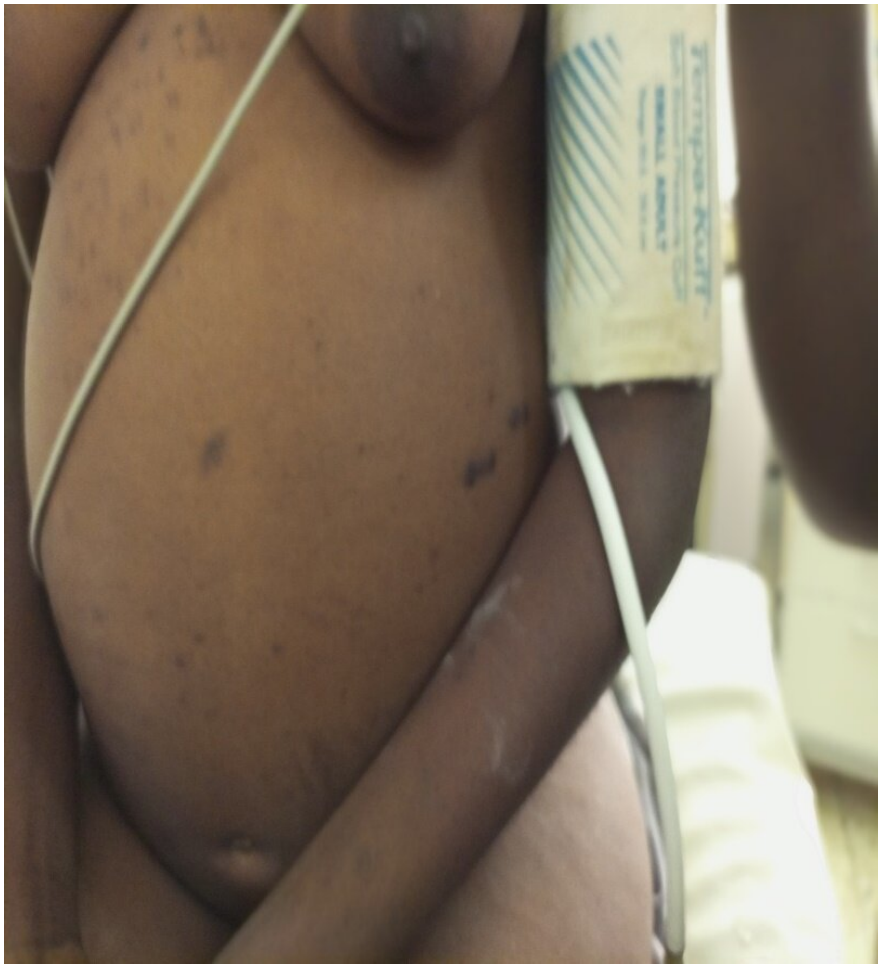
Figure 1:Todani classification of choledochal cyst

Case presentation:

A 17 year old female child who presented who presented with 4 months history of constant, dull, non-radiating abdominal pain and a progressively increasing right upper quadrant mass. This was associated with yellowish discoloration of eyes, urine and clay colored stools. Reported one episode of vomiting but no history of nausea, diarrhea, fever or recent weight loss. There was no significant past medical or surgical history. One month prior to admission, she reported to had visited a clinic in Ibanda district where she was told about referral to a bigger facility for definitive management but due to financial constraints, she wasn't able to come to referral facility in time.

Clinical findings:

On physical examination:



She was in fair general condition, fairly well nourished, with obvious deep jaundice with a big mass extending from the epigastrium to the hypogastrium. The mass was non tender, soft, non-mobile, couldn't go above it and below it, measuring 30cm x 20cm (Figure 2). There were no visible collateral vessels, no inguinal masses or palpable lymph nodes

Diagnostic assessments:

Blood work up included complete blood count, serum electrolytes, liver function tests, renal function tests, INR, RCT and Hepatitis surface antigens (Table 1). CT scan was done and it revealed a huge cystic mass in the right upper quadrant extending inferiorly to the right lumbar region, surrounded by a thin enhancing wall on intravenous contrast. It was continuous with common bile duct and was compressing the gall bladder, head of pancreas and the right kidney (Figure 3). There was no focal or diffuse masses seen on the liver and there was no regional lymphadenopathy.

Table 2: Laboratory tests showing pre-operative and post-operative results

Parameter Blood	**Pre- Operative Laboratory values**	**Post- Operative Laboratory values**	**Reference Range of MRRH Laboratory**
White blood count(10ˆ9/L)	6.7	5.5	4.00-10.00
Haemoglobin(g/dL)	10.0	12.0	11.0-15.0

Parameter Blood	**Pre- Operative Laboratory values**	**Post- Operative Laboratory values**	**Reference Range of MRRH Laboratory**
Platelets(10ˆ9/L)	408	390	150-450
Bilirubin-total(mg/dL)	6.41	0.9	0.1-1
Bilirubin-direct(mg/dL)	4.8	0.17	0-0.2
Alanine Transaminase(u/l)	144	34	0-41
Aspartate Transaminase(u/l)	208	39	13-40
Alkaline phosphatase(u/l)	1827	1190	0-1200
Total protein(g/dL)	9.35	7.0	10-15
Pro-thrombin time(sec)	18	14	10-15
International Normalized Ratio(INR)*	1.72	1.1	0.8-1.2
CA 19-9*	12	13	0-36
CEA(ng/mL)*	1.2	1.2	<2.5
Hepatitis B surface antigens	Negative	Not repeated	
HIV serology	Negative	Not repeated	
Urinalysis Proteins Leukocytes Bilirubin	proteinuria++ + ++	Not repeated	

Note :* Investigations are not available at MRRH Laboratory, therefore were done from the private laboratory

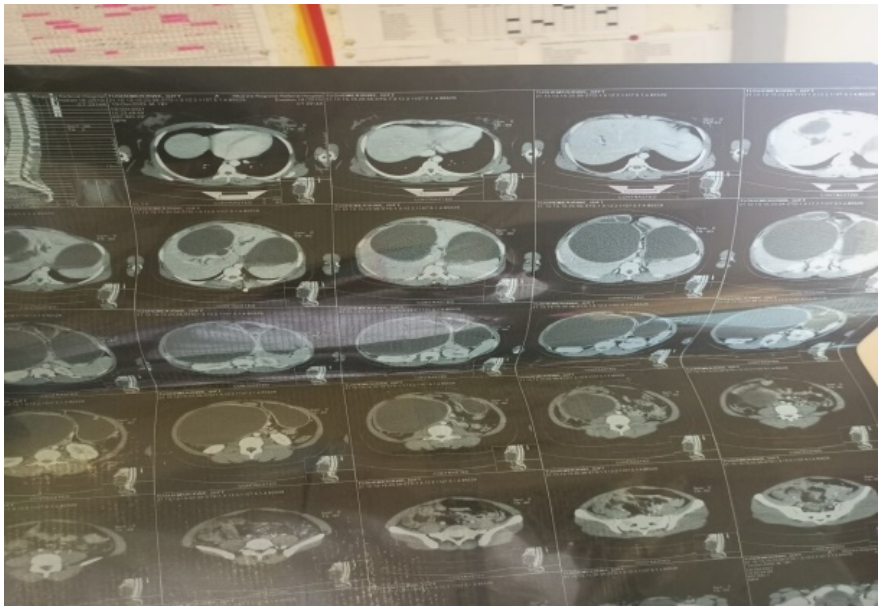


Figure 3: CT scan findings (CT scan showing a huge thin-walled, non-enhancing cystic mass rising from the right hypochondrium to the right lumbar region)

Therapeutic intervention:

Laparotomy through an extended midline incision was done. A gross enlarged cystic mass extending from the liver up to the hypo-gastric region with the duodenum, the gall bladder, greater omentum and transverse colon adherent onto the cystic mass (Figure 4). Adhesions were careful divided, separating the duodenum, transverse colon from the cystic mass, and achieved hemostasis. Dissection continued medially and laterally to release the attachments of the cyst from other intra-abdominal wall structures. The mass was approximately 30cmx 25cmx 20cm containing around 3 liters of greenish, bilious fluid (Figure 5 & 6). Cystic duct was identified and anterograde cholecystectomy was performed. A loop of jejunum 45cm from the ligament of treitz was brought near the stump of common hepatic duct through a defect created on the left side of the middle colic vessels in the transverse mesocolon. Hepaticojejunostomy and jejunojejunostomy was done using Braun procedure (Figure 7). A drain was left in proximity to the site of biliary reconstruction and was removed 6 days later after spending 3 days without any collection in its bag. The specimen was sent for histo-pathological examination and was reported to contain chronic inflammatory infiltrate with no evidence of malignancy and the patient recovered.

Follow- up and outcomes:

Two months of the review, patient was all normal both clinically and on blood workup

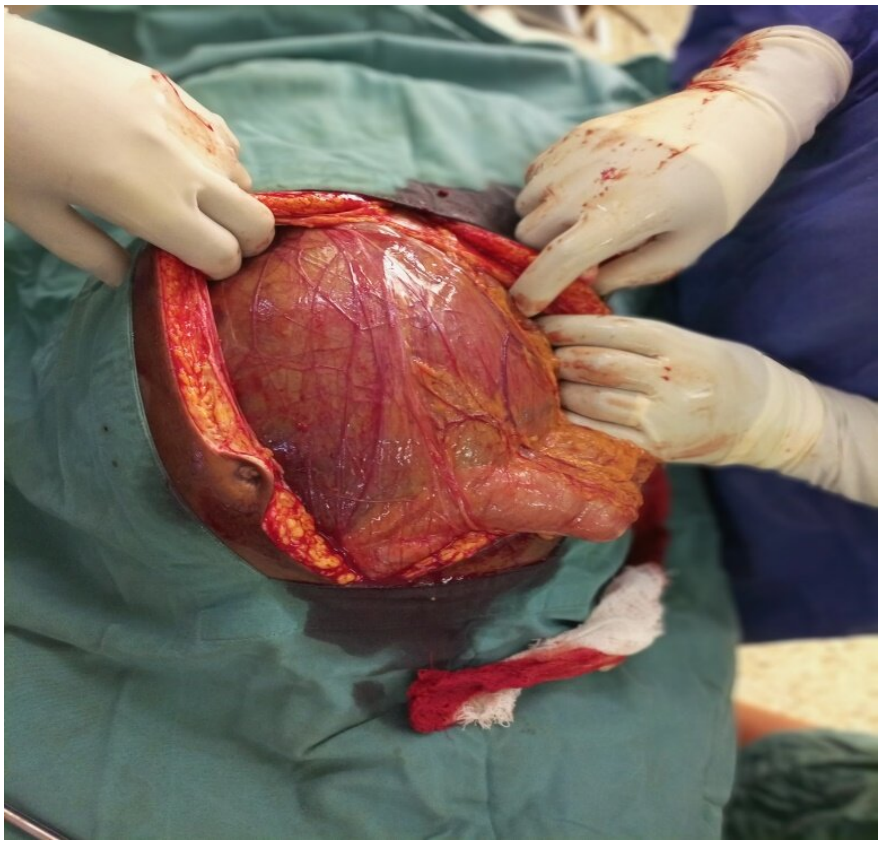


Figure 4: Intraoperative view showing part of transverse colon adherent to the cyst

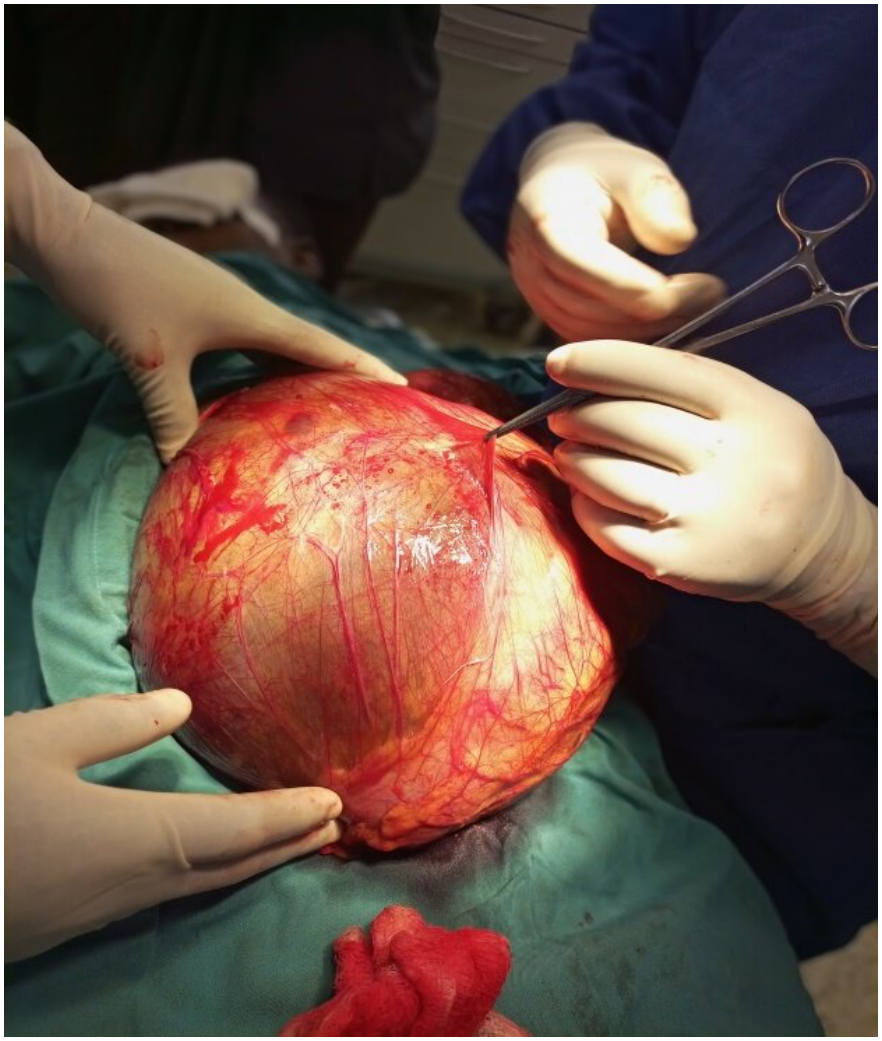


Figure 5: Intraoperative view showing giant choledochal cyst

Figure 6: Shows the greenish bilious content drained from a huge choledochal cyst which was approximately 5 liters

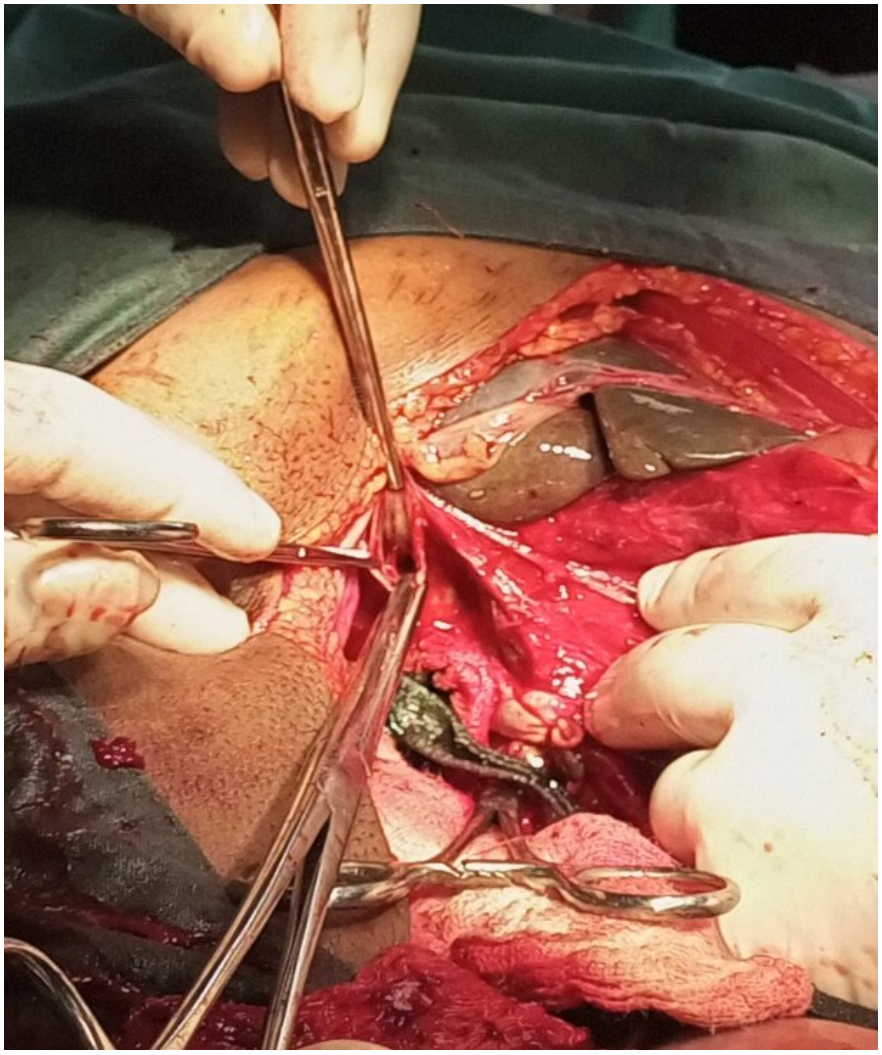


Figure 7: Shows right and left hepatic ducts where the common hepatic was used in the hepaticojejunal anastomosis

Discussion:

Choledochal cysts are infrequent congenital malformations characterized by cystic dilatations of the biliary tree(1). These dilatations can be extra-hepatic, intrahepatic or both. The incidence of choledochal cysts in Asian population is 1 in 1000 live births with predominance in females. In western countries, the incidence ranges from 1:100,000 to 1:150,000 in the United States, 1 in 2 million in the United Kingdom(12). In Uganda, so far this is the second case of giant choledochal cyst in a young female adult that has been managed successful at Mbarara Regional Referral Hospital (13). A study done in Uganda by Odongo et al., shows that choledochal cysts account for 1.4% of the causes of extra-hepatic obstructive jaundice which is very low hence making it rare case(14)

Giant choledochal cyst refer to the huge cysts with a diameter of greater than 10 cm(15). For our case, intra-operatively, the cyst was measuring 30cm x 25cm x 25cm which is big.

The primary diagnostic modality is by abdominal ultrasound scan followed by CT scan and the MRI(16). The associated anomalies are biliary atresia, gallbladder atresia, hepatic fibrosis and those of pancreatico-

biliary ductal system. MRI and MRCP have a conclusive role in confirming ultrasound scan diagnosis(5). This is also done to evaluate anatomy and identify complications related to the cyst(5). In our case, only imaging modalities available were abdominal ultrasound scan and the CT-scan. The abdominal ultrasound sound scan was used to screen the patient and identify the choledocal cyst. The CT-Scan was used to confirm our diagnosis and classify our cyst under Tonadi classification and it was type IA choledochal cyst.

Although studies have documented the use of ERCP in certain centers of Uganda in the management of hepatobiliary conditions, in our center (MRRH), there is absence of these modern invasive machines that can help us in both diagnosis and treatment of choledochal cysts. Therefore we rely on open surgery for everything which in some instances increases the chances of complications for example along hospital stay(17).

The treatment of choledochal cyst is an initial control of complications then followed by the primary surgery of the cause. Presently the most popular and accepted surgical intervention is by total excision of the choledochal cyst and restore the biliary enteric drainage into the duodenum either via Roux-en-Y hepatico-jejunostomy or Braun hepatico-jejunostomy(18). The later approach was used in our patient to reconstruct a connection between common hepatic duct and a loop of jejunum 40 cm from the ligament of treitz(10).

Conclusion

Giant choledochal cysts is a very rare disease which can be diagnosed using abdominal CT scanning since it offers a great deal of information that is helpful in both diagnosis and planning for surgical approaches . Abdominal ultrasound scan is the initial screening in patients with choledochal cysts and can also be used to make diagnosis antenatal. Use of MRI and MRCP techniques is increasing for the diagnosis of choledochal cysts however they are not easily available and is expensive. Complete surgical excision is very important since it reduces the chances of pancreatitis and biliary malignant transformation.

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Ethical Approval:

Ethical approval was obtained from Mbarara University of Science and Technology Research Ethics Committee (REC)

Consent:

Written assent was obtained from the patient's guardian for publication of this case report and accompanying images and is available for the review by the Editor-in-Chief of this journal on request.

Author contributions:

All authors made an important contribution to the work reported, whether that is in the concept design, study design, acquisition of data, analysis and interpretation of data; took part in drafting, revising or reviewing of the article carefully; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be held responsible for all aspects of this article

Disclosure

The authors report no conflicts of interest in this work.

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