

Choledochal Cyst-Current Management Strategies

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Abstract: Choledochal cysts in adults are often detected during the investigation of nonspecific symptoms. Choledochal cysts are uncommon and of unknown cause, with an incidence of 0.1% even among adults referred for endoscopic retrograde cholangiopancreatography (ERCP) investigation. There is a higher incidence in females. The presentation is often vague and nonspecific, impeding prompt diagnosis. However, the diagnosis is facilitated by modern imaging techniques. While some aspects of optimal management have been resolved, others remain controversial. Here in we report our experience concentrating particularly on current issues in management of the disease.

MRCP is the best imaging modality for diagnosis and characterization of these cysts. Complete excision and hepaticojejunostomy is the management of choice. Two drains (Subhepatic and pelvis) should be put at time of surgery.

Keywords: Choledochal cysts, ERCP, hepaticojejunostomy

INTRODUCTION

Choledochal cysts are uncommon and of unknown cause, with an incidence of 0.1% even among adults referred for endoscopic retrograde cholangiopancreatography (ERCP) investigation. There is a higher incidence in females. The presentation is often vague and nonspecific, impeding prompt diagnosis. However, the diagnosis is facilitated by modern imaging techniques. While some aspects of optimal management have been resolved, others remain controversial. Here in we report our experience concentrating particularly on current issues in management of the disease.

MATERIAL AND METHODS

Experience of 12 cases of Choledochal cyst- males 3, females 9, M:F 1:3, mean age -21 years, Incidental with vague symptoms -6, stones in choledochal cyst -2, Cholangitis-4. All vague symptoms cases were initially diagnosed by USG out of which 2 were confirmed by ERCP and rest 4 by MRCP. Stones in choledochal cyst (2) suspected by USG and confirmed by ERCP. All four cholangitis presentation cases were suspected by USG and confirmed by MRCP (table 1). Complete excision of cyst was possible in 7 cases, while 3 cases upper cuff left for anastomosis and in two cases lower portion mucosa fulgurated and left in situ. No intraductal drain left, only drains in Morrison's pouch and pelvis left. 3 had minor leak with drain out put erris not more than 1 week, while 1 patient had a leak with drain out put varying from 400-600 ml daily during first 4 weeks and then 200-250 ml daily during next 2 weeks and then on 49th day no discharge in bag, Pelvic drain helped in all these 4 leak patients and waste removed from 7th to 10th day.

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Table 1

S. no.	Presentation	No. of Cases	Initially Suspected	Diagnosis by
1	Vague Symptoms	2 4	USG USG	ERCP MRCP
2	Stone Choledochal cyst	2	USG	ERCP
3	Cholangitis	4	USG	MRCP

DISCUSSION

Choledochal cysts are rare abnormalities of biliary tree and so may be frequently overlooked in differential diagnosis. The nonspecific symptoms of choledochal cysts, including pain in upper abdomen and jaundice, are common in many other illnesses of upper gastrointestinal tract. The clinical triad of jaundice, right upper quadrant mass and abdominal pain occurs only in minority of patients (0%-17%) (3,5,6,7). It is more commonly seen in cases with onset in childhood rather than in adults, and 85% of children have at least 2 features of the triad at presentation, compared with only 25% of adults (8).

Among the more commonly reported presenting features are cholangitis, (6, 9, 10) pancreatitis, (6, 10, 11, 12) and biliary peritonitis from cyst rupture (6, 13, 14, 15). Among adults, choledochal cysts are quite often incidental findings during investigation for other problems (6, 16, 17, 18). The most commonly used classification developed by Alonso-Lej et al (1959)¹⁹ and modified by Todani et al (1977)²⁰ describes 5 broad types of choledochal cysts. However, it is also not without controversy. Some have argued that the term "Choledochal cyst" should refer to only type I and IV cysts (which comprise over 90% of biliary cysts) Vague abdominal pain is the prominent complaint in adults, which led all of our patients to seek medical attention, hence high index of suspicion should be observed in adult patients to have a prompt diagnosis. In adults, the diagnosis is only sporadically in doubt preoperatively, owing to the quality of modern imaging (1). Ultrasonography is usually the first examination and is very sensitive in the detection of cystic structures. A computed tomography usually can give more information and modern techniques, including reconstruction, should allow for establishing the diagnosis. A better role for CT scanning may be in the postoperative period, where it was shown to be superior to MRCP in locating the biliary enteric anastomosis and in defining any stenosis there of (1).

Endoscopic retrograde cholangiopancreatography (ERCP) although invasive can precisely visualize the extrahepatic bile duct (helped in confirming diagnosis in two patients presented with stone in cystic duct and rest 2 patients were from vague symptoms group prior to availability of MRCP). MRCP is currently the preferred reliable investigation as it also precisely visualizes complete biliary tree with advantage of being noninvasive MRCP helped in two third (8/12) of our cases.

Pancreatitis, cholangitis, Biliary cirrhosis and malignancy are the complications reported in choledochal cysts (1).

The treatment of choledochal cyst has changed. In the past, a cystojejunostomy was the standard procedure. Currently, excision of the cyst and reconstruction by hepaticojejunostomy is the standard therapy (1). The timing of surgery should be early after diagnosis to reduce the incidence of complications (22).

Postoperative anastomotic structure formation may be reduced by performing a higher anastomosis, as Todani (23) found that 9 of 22 choledochojejunostomies had postoperative anastomotic strictures, compared with 1 of 82 hepaticojejunostomies.

Complete excision of cyst was possible in 58% (7/12) cases, while 25% (3/12) upper cuff of cyst was left for anastomosis with Jejunum as right and left hepatic ducts were 23 mm in diameter. In 16.6% (2/12) cases lower part of cyst in pancreatic head area fulgurated and left in situ.

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25% (3/12) cases had minor leak with drain out put persisting for 4-6 days. Pelvic drain helped in these leak cases as it drained in 1st post operative week and removed on 7th to 10th day. One of our case had bile leak 400-600 ml per day for almost a month and then 200-250 ml daily for next 2 weeks on 49th day the drain stopped draining. This was probably because of small bile duct directly draining from liver into GB (Ducts of Luschka) unrecognized at time of surgery and hence not ligated.

SUMMARY

Choledochal cysts in adults are often detected during the investigation of nonspecific symptoms. MRCP is the best imaging modality for diagnosis and characterization of these cysts. Complete excision and hepaticojejunostomy is the management of choice. Two drain (Subhepatic and pelvis) should be put at time of surgery.

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