Pediatr Surg Int (2013) 29:777–786
DOI 10.1007/s00383-013-3333-5

ORIGINAL ARTICLE

Variations in biliary ductal and hepatic vascular anatomy and their relevance to the surgical management of choledochal cysts

Richa Lal · Anu Behari · Ranjit Hari Vijaya Hari · Sadiq S. Sikora · Surender Kumar Yachha · Vinay Kumar Kapoor

Accepted: 11 June 2013 / Published online: 22 June 2013

Abstract
Purpose An aberrant biliary ductal and vascular anatomy presents a technical challenge for choledochal cyst (CDC) surgery. Mismanagement may have unfavourable implications. This study highlights the spectrum, approach to their identification and management.
Methods Forty of 117 (34 %) cases were identified to have an aberrant biliary ductal ($n = 17$) or arterial ($n = 26$) anatomy; 3 had both. The pancreaticobiliary anatomy was defined by an intraoperative cholangiogram (IOC) before January 2005 and a preoperative magnetic resonance cholangiopancreatogram (MRCP) subsequently.
Results IOC missed 3 of 4 aberrant biliary ducts, while an MRCP accurately delineated 10 of 13 aberrant bile ducts. The significant biliary anomalies were: an aberrant right sectoral/segmental duct joining the common hepatic duct (CHD) or the cyst itself ($n = 14$), cystic duct ($n = 1$) and cystic duct–CHD junction ($n = 1$). The aberrant duct was incorporated into the biliary-enteric anastomosis (B-EA) by: (i) double ostia B-EA ($n = 1$), (ii) ductoplasty with single ostium B-EA for aberrant duct and CHD ($n = 2$), and (iii) transection of the CHD/cyst distal to the aberrant duct orifice with a single ostium B-EA ($n = 13$). The arterial anomalies were (i) replaced or accessory right hepatic artery (RHA) ($n = 11$) and (ii) RHA crossing anterior to the cyst ($n = 15$), which was repositioned posterior to the B-EA.
Conclusion It is important to consciously look for, appropriately identify and manage aberrant biliovascular anatomy. MRCP facilitates accurate preoperative delineation of aberrant duct anatomy. All major aberrant ducts need to be incorporated into the B-EA and aberrant arteries should not be ligated.

Keywords Choledochal cyst · Congenital choledochal malformation · Biliary anatomy · Aberrant bile duct · Arterial anomaly · Magnetic resonance cholangiopancreatography

Introduction

The standard treatment of a choledochal cyst, also referred to as congenital choledochal malformation, is excision of the extrahepatic cyst and a wide, hilar, biliary-enteric anastomosis (B-EA). Biliary ductal and vascular anomalies at the hepatic hilum present a technical challenge in the surgery of choledochal cysts.

Anatomical variations are encountered so frequently that it is often said that anatomic variability is a rule rather than an exception in liver and biliary surgery [1].

Awareness of anatomical variations is important because inability to recognize and appropriately manage biliary ductal and vascular anomaly can lead to serious intra and postoperative complications in resective and reconstructive surgery of the biliary tree at the hepatic hilum [1].

Understanding of biliary ductal and vascular anatomy and its variations has improved significantly as experience

R. Lal (✉)
Department of Pediatric Surgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow 226014, India
e-mail: richalal@gmail.com

A. Behari · R. H. V. Hari · S. S. Sikora · V. K. Kapoor
Department of Surgical Gastroenterology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow 226014, India

S. K. Yachha
Department of Pediatric Gastroenterology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow 226014, India

with living related and split liver transplantations has grown. Most surgical and radiological studies in literature have highlighted the significance of recognizing aberrant anatomy in harvesting donor liver grafts in living related liver transplants and split liver transplants and during laparoscopic cholecystectomy so as to avoid inadvertent injury and perform appropriate reconstructive procedure safely [1–6].

However, there is scant literature on aberrant arterial and biliary ductal anatomy with reference to surgery for choledochal cysts.

This article discusses the normal and anomalous hepatic vascular and biliary ductal anatomy, its identification and appropriate management as is relevant to the surgical management of choledochal cysts based on our experience of 117 cases of choledochal cysts operated from February 2000 to May 2012.

Methods

Between February 2000 and May 2012, 117 children (age: 40 days–14 years) with choledochal cysts were managed at a tertiary care referral hospital in North India. The clinical presentation, radiological findings, operation details with special reference to arterial and biliary ductal anatomy and the outcome after surgery were analyzed from a prospectively maintained database.

Before January 2005, the radiological imaging studies used for diagnosis and delineating the precise pancreaticobiliary anatomy were an abdominal ultrasonogram followed by an intraoperative cholangiogram ($n = 32$). The intraoperative cholangiogram was performed by cannulating the cystic duct with a 5-Fr infant feeding tube or a 4-Fr ureteric catheter, the tip of which was advanced into the common bile duct distally. The gall bladder end of the cystic duct was ligated to prevent dye from getting sequestrated into the gall bladder. After January 2005, ultrasonogram followed by a preoperative magnetic resonance cholangiopancreatogram (MRCP) ($n = 82$ cases) became a part of routine preoperative diagnostic protocol. However, MRCP was supplemented with an intraoperative cholangiogram if the image quality of preoperative MRCP was suboptimal, especially in infants or if the pancreaticobiliary duct junction was not clearly visualized on MRCP. MRCP was performed on a 3-T scanner (Signa HDxT®, GE Healthcare, Milwaukee,Wisconsin, USA) using a 8US TORSOPA coil for data acquisition. After localizer sequence, axial T2-weighted SSFSE sequence was done with respiratory gating. MRCP images were acquired in coronal and coronal oblique planes by heavily T2-weighted sequence with the following image parameters: effective echo time 792.4 ms, repetition time 4,249 ms, slice thickness 40–50 mm. A respiratory gated coronal 3D MRCP sequence was also done with the following parameters: effective echo time 663.9 ms, repetition time 2,609 ms, slice thickness 0.8 mm with 0.4 mm overlap, matrix 320 × 256/0.75 NEX. Patients were given oral gadodiamide (Omniscan, Amersham health, cork, Ireland) solution diluted 1:15 in water just before starting the study to suppress signal intensity of other fluid-containing organs. The best imaging plane was selected on the basis of visualization of the pancreaticobiliary system.

The other modalities for obtaining a preoperative cholangiogram were a T-tube cholangiogram ($n = 1$) and a cholangiogram done by injecting dye via a percutaneous transhepatic biliary drainage (PTBD) catheter ($n = 2$). The T-tube cholangiogram and a PTBDgram were done in the radiology suite under fluoroscopy. The indication for T-tube placement was presentation as spontaneous perforation with biliary peritonitis and for a preoperative PTBD catheter severe cholangitis recalcitrant to intravenous antibiotic therapy.

A conscious effort was made to define sectoral and segmental biliary ductal anatomy precisely on preoperative or intraoperative cholangiogram and to correlate it with the operative findings.

On the basis of the cholangiogram and operative findings, the extrahepatic biliary ductal anatomy has been classified into six main types [7] and the anatomical variations together with their reported frequency are as follows [3, 7]. In type A (57 %), the anatomy is normal. In type B (12 %), there is trifurcation of the common bile duct into right anterior, right posterior and left hepatic duct with an absence of a demonstrable length of the right hepatic duct. In type C (20 %), there is an aberrant drainage of the right segmental ducts into the common hepatic ducts. In type D (6 %), there is an aberrant drainage of the right segmental duct into the left hepatic duct. In type E (3 %), there is absence of hepatic duct confluence, with convergence of two or more ducts from either lobe to form the common hepatic duct. Finally, in type F (2 %), there is an absence of the right hepatic duct with ectopic drainage of the right posterior duct into the cystic duct.

In addition, a note was made of the anomalies related to variations of cystic duct anatomy which would have an implication on the surgical technique [7].

Similarly, a conscious effort was made to visualize and palpate arterial pulsations anterior and postero-lateral to the choledochal cyst before encircling and transecting it. The anomalous arteries could be accessory, occurring in addition to the normal arterial supply, or replaced, representing the primary arterial supply to the lobe. The arterial anatomy variations were classified as follows as defined by Hiatt et al. [8] based on a study on 1,000 cases that underwent donor liver harvesting for live-related liver transplantation:

Type 1: in this normal pattern, the common hepatic artery arises from the celiac axis to form the gastroduodenal and proper hepatic arteries; the proper hepatic artery divides distally into the right and left branches.
Type 2: a replaced or accessory left hepatic artery arises from the left gastric artery.
Type 3: a replaced or accessory right hepatic artery originates from the superior mesenteric artery.
Type 4: a double replaced pattern; the right hepatic artery arises from the superior mesenteric artery and the left hepatic artery as a branch of the left gastric artery.
Type 5: the entire common hepatic artery originates as a branch of the superior mesenteric artery.
Type 6: the common hepatic artery takes direct origin from the aorta.

In addition, the hepatic artery proper may cross anterior to the bile duct in as many as 20 % of cases [7].

The standard definitive surgical procedure performed was excision of the extrahepatic part of the choledochal cyst with a Roux-en-Y hepaticojejunostomy using a 40-cm retrocolic jejunal Roux-en-Y loop, while ensuring a wide, hilar, biliary-enteric anastomosis (B-EA). The B-EA was carried proximal to the confluence onto the extrahepatic part of the left and/or right duct if the cholangiogram and the operative findings suggested a primary ductal stenosis at or proximal to the confluence.

After having encircled the choledochal cyst or the common hepatic duct (CHD) at the appropriate level, the anterior wall of the duct/cyst was initially incised distal to the confluence and the lumen was carefully inspected from inside for any aberrant duct orifice distal to the confluence which might have been missed on preoperative imaging. Circumferential duct transection was subsequently carried out at a more proximal level if necessary. This simple extra precautionary step prevented an inadvertent injury to an aberrant duct inserting onto the CHD or the choledochal cyst in cases wherein the aberrant duct was missed on a pre- or intraoperative cholangiogram.

All aberrant ducts were incorporated into the B-EA and none were ligated. The technique of incorporation of the aberrant duct into the B-EA was determined by the diameter of the aberrant duct and its distance from the confluence. The various surgical approaches employed were as follows—(i) a double ostium B-EA (Fig. 1): this was indicated for a large-diameter aberrant duct located far from the confluence and, hence, could not be approximated with the CHD at the confluence with ease. The aberrant duct and the CHD were implanted onto the Roux-en-y loop of the jejunum separately. (ii) Ductoplasty with a single ostium B-EA: this was indicated for a small-diameter aberrant duct inserting onto the cystic duct or cystic duct–CHD junction (Fig. 2a) and which could be approximated

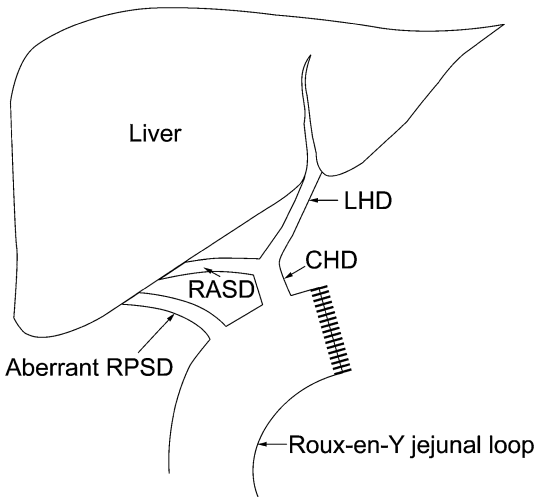

Fig. 1 Double ostia biliary-enteric anastomosis: the confluence is formed by the junction of the left hepatic duct (LHD) and right anterior sectoral duct (RASD). The aberrant right posterior sectoral duct (RPSD) and the common hepatic duct (CHD) were implanted onto the Roux-en-Y jejunal loop separately

with the CHD at the confluence without tension. The aberrant duct was approximated with the CHD at the confluence with interrupted fine 5-0 polydioxanone sutures (PDS). The aberrant duct orifice was enlarged by making an incision along its lateral wall followed by a single ostium B-EA (Fig. 2b). (iii) Transection of the CHD/cyst distal to the aberrant duct orifice with a single ostium B-EA (Fig. 3): this was indicated for a small-diameter aberrant duct joining the CHD/cyst close to the confluence. Here, the CHD/cyst was transected distal to the aberrant duct orifice, thereby retaining a thin rim of the cyst/CHD between the aberrant duct and the confluence, followed by a single ostium B-EA.

The postoperative follow-up protocol consisted of the following—(i) symptoms: pain, jaundice, cholangitis and pancreatitis; (ii) liver function test : especially serum conjugated bilirubin and serum gamma glutamyl transpeptidase (GGT)]; (iii) ultrasonography: to assess for persistence or progressive regression of intrahepatic bile duct dilatation [9] or hepaticolithiasis; (iv) radio isotopic Tc^{99m} mebrofenin hepatobiliary scintigraphy: a satisfactory drainage across the B-EA was defined as appearance of the radio-isotope in the bowel loops within 20 min of radio-isotope injection followed by total clearance from the liver parenchyma, intrahepatic bile ducts and anastomotic site by 1 h in a patient with normally functioning liver parenchyma [10]. These were performed at six monthly intervals in the first 2 years following surgery, followed by a yearly follow-up. Postoperative MRCP was indicated only if the above-mentioned evaluation suggested a possibility of obstruction at the B-EA. However, in clinical decision-making, a combination of parameters was taken into

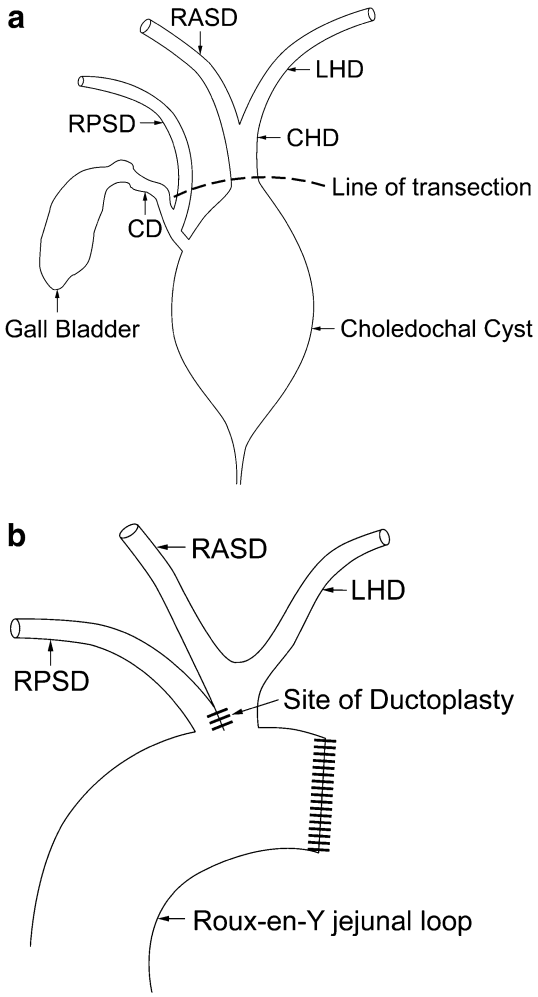

Fig. 2 **a** Aberrant right posterior sectoral duct (RPSD) inserting into the cystic duct (CD). The confluence is formed by the joining of the left hepatic duct (LHD) and right anterior sectoral duct (RASD). The *dotted line* of transection depicts the level at which the CHD and the aberrant RPSD would be divided; **b** ductoplasty and a single ostium biliary-enteric anastomosis: the aberrant right posterior sectoral duct (RPSD) which was disconnected from its cystic duct insertion was approximated with the common hepatic duct (CHD) at the confluence with fine interrupted sutures. A single ostium B-EA was made

consideration. Also, any persistent abnormality, rather than a single isolated deranged report, was considered to be of significance.

Long-term outcome was assessed by a combination of the above-mentioned parameters.

Results

The study population comprised 117 consecutive patients (age range 40 days–14 years) treated surgically for choledochal cysts between February 2000 and May 2012.

The commonest definitive surgical procedure performed was excision of the extrahepatic choledochal cyst and

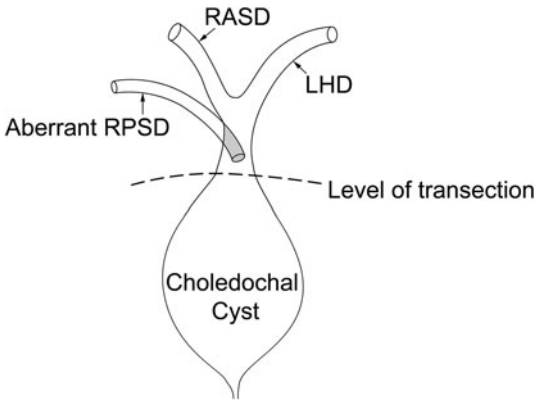

Fig. 3 Aberrant RPSD is inserted into the relatively normal-caliber CHD proximal to the cystically dilated CBD. The *dotted line* of transection indicates that the CHD is divided distal to the orifice of the aberrant right posterior sectoral duct (RPSD) followed by a single ostium B-EA

Roux-en-Y hepaticojejunostomy ($n = 114$). Seven of these patients were referred to us after a prior surgical intervention in the form of T-tube drainage of the choledochal cyst for spontaneous perforation and biliary peritonitis ($n = 2$); an internal drainage procedure ($n = 3$); and simple primary closure for spontaneous cyst perforation and biliary peritonitis ($n = 2$). For each of these patients, an elective cyst excision and Roux-en-Y hepaticojejunostomy were performed at our center. In addition, revision of a strictured hepaticojejunostomy was done for one patient who was referred to us with recurrent cholangitis. Two infants, 9 months of age each, underwent a cystojejunostomy and a tube hepaticostomy, respectively, because secondary biliary cirrhosis, decompensated liver disease and extensive collaterals in the hepatoduodenal ligament precluded a safe cyst excision in each of them.

Of these 117 cases, 17 (14.5 %) were identified to have a biliary ductal anomaly, 26 (22 %) had an arterial anomaly and 3 had co-existent arterial and biliary ductal anomaly, each of which had an implication on the surgical technique.

Sixteen of 40 cases (40 %) with aberrant bilio-vascular anatomy had varying grades of intrahepatic duct dilatation preoperatively.

From the management perspective, intrahepatic duct dilatation was differentiated into two anatomical groups based on cholangiography and intraoperative findings [11].

Group 1 ($n = 11$) comprised cases with contiguous upstream intrahepatic duct dilatation secondary to raised intracystic pressure. A wide B-EA made at or distal to the confluence resulted in progressive regression of upstream dilatation.

Group 2 ($n = 5$) cases had disproportionate dilatation of the left and/or right duct as compared to the common hepatic duct with primary ductal stenosis at the confluence

or proximal to it [11]. The sites of stenosis were as follows: (i) stenosis at the left duct orifice ($n = 2$); (ii) stenosis at both the left and right duct orifice ($n = 1$); (iii) septum at the confluence ($n = 1$); (iv) stenosis at the aberrant duct orifice ($n = 1$). In each of these cases, the B-EA was carried proximal to the site of stenosis onto the extrahepatic part of the left, right or the aberrant duct with/without excision of the membranous web or septum if present. Incidentally, all five cases in this group were associated with aberrant arterial anatomy.

Aberrant biliary ductal anatomy: identification, types and management

Preoperative MRCP and intraoperative cholangiograms: their role in detection and precise delineation of the biliary ductal anatomy

Before the advent of routine preoperative MRCPs in the current study, an intraoperative cholangiogram ($n = 32$) detected a biliary ductal anomaly in only one case. In three other cases, an intraoperative cholangiogram missed an aberrant duct which was incidentally detected during cyst dissection and transection.

After January 2005, as MRCPs became a part of routine preoperative imaging protocol ($n = 82$), the presence of an aberrant duct was accurately delineated on preoperative MRCP in 10 of the 13 cases which were eventually demonstrated to have an aberrant duct at surgery. MRCP also precisely defined the liver sector or segment drained by each of these aberrant ducts and the site of insertion of the aberrant duct onto the CHD, the cyst itself or the cystic duct. In one of these ten cases, the clue to the presence of an aberrant duct was the inability to visualize the posterior sectoral duct at the confluence. A conscious effort to localize an aberrant duct revealed an undilated aberrant right posterior sectoral duct visualized only as a faint hair-like structure joining the CHD–cystic duct junction. This was documented more clearly on an intraoperative cholangiogram, wherein the contrast was injected under some pressure.

However, aberrant ducts were missed on preoperative MRCP in 3 of 13 cases. These were detected incidentally at surgery.

Precise anatomy of aberrant ducts identified ($n = 17$)

Seventeen of the 117 cases (14.5 %) had a biliary ductal anomaly which had an implication on the surgical dissection, cyst excision and reconstruction. They were as follows: (i) aberrant opening of the right sectoral duct (anterior or posterior) into the relatively normal-caliber CHD proximal to the cystically dilated common bile duct (CBD) (Figs. 3, 4a) or the cyst itself (Fig. 4b) (type C variation [7] in biliary ductal anatomy) ($n = 14$); (ii) ectopic opening of the right sectoral duct into the cystic duct (Fig. 2a) (type F variation [7] in biliary ductal anatomy) ($n = 1$); (iii) aberrant opening of the right sectoral duct at the cystic duct–CHD junction ($n = 1$). In addition, the cystic duct was observed to join the right anterior sectoral duct in one case. In each of these locations, the aberrant duct, if unrecognized, is at risk of injury during duct/cyst transection or during cholecystectomy. Patients with type B, D and E variations in biliary ductal anatomy [7] as defined in "Methods" were excluded from the study because this anatomy did not have any implication on the surgical technique of excision and reconstruction of choledochal cyst.

Further anatomical details of major ductal anomalies in terms of liver segments drained, the site of insertion of the aberrant duct on extrahepatic biliary tree, the anatomical cyst type as per Todani's classification [12] and the associated anomalies have been summarized in Table 1.

Management of aberrant bile ducts

All aberrant ducts were incorporated into the B-EA and none were ligated. The approach to incorporation of an aberrant duct into the B-EA was governed by the diameter of the aberrant duct and its distance from the confluence as mentioned in "Methods". Accordingly, the surgical techniques employed were as follows:

(i) Double ostia B-EA ($n = 1$) (Fig. 1).
(ii) Ductoplasty with single ostium B-EA ($n = 2$) (Fig. 2b).
(iii) Transection of the CHD or the cyst distal to aberrant duct orifice with a single ostium B-EA ($n = 13$): Here, a thin rim of the cyst or the CHD between the aberrant duct orifice and the confluence is retained (Fig. 3). In 8 of these 13 cases, the aberrant duct was inserted onto a relatively normal-caliber CHD proximal to the cystically dilated CBD (Table 1). Hence, this approach did not leave behind residual disease in these eight cases.
(iv) The cystic duct joining the right anterior sectoral duct was carefully dissected along its course and the junction of the cystic duct with the right anterior sectoral duct carefully repaired.

Variations of arterial anatomy: types and management

Types of aberrant arterial anatomy identified:

Twenty-six of 122 cases (21 %) were identified to have a variation in the arterial anatomy which had an implication on the dissection and reconstruction. These were:

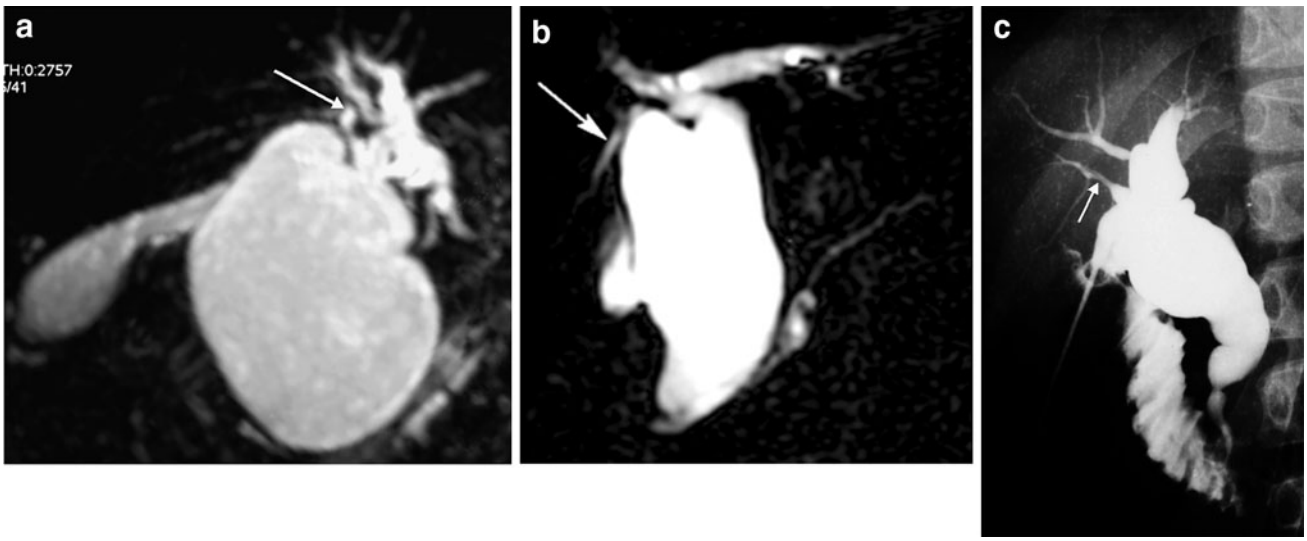

Fig. 4 **a** *Arrow* indicates an aberrant segment five duct inserted onto the relatively normal-caliber CHD proximal to the cystically dilated CBD; **b** *arrow* indicates an aberrant right sectoral duct inserted onto a Type 1a cyst itself; **c** *arrow* indicates an aberrant right sectoral duct inserted onto a Type 1c cyst. The duct was faintly visualized on MRCP, but was more apparent on direct contrast injection intraoperative cholangiography

Table 1 Anatomical details of major aberrant bile ducts identified ($n = 17$)

Sectoral/segmental anatomy of the aberrant duct (number of cases)	Site of insertion on the extrahepatic biliary tree	Cyst type (Todani's classification [12])	Associated anomaly
Right posterior sectoral duct ($n = 3$) Right anterior sectoral duct ($n = 3$) Segment five duct ($n = 2$)	Common hepatic duct (CHD) proximal to the cystically dilated common bile duct (CBD) ($n = 8$) (Figs. 3, 4a)	Type 1a	Right hepatic artery crossing anterior to the CHD ($n = 1$)
Right anterior sectoral duct ($n = 1$) Right sectoral duct* ($n = 2$)	Cyst itself ($n = 3$) (Fig. 4b)	Type 1a	Accessory right hepatic artery ($n = 1$)
Right posterior sectoral duct ($n = 2$) Right anterior sectoral duct ($n = 1$)	CHD dilated in fusiform configuration ($n = 3$) (Fig. 4c)	Type 1c	Right hepatic artery crossing anterior to the CHD ($n = 1$)
Right anterior sectoral duct ($n = 1$)	Cystic duct–CBD junction	Type 1a	None
Right sectoral duct* ($n = 1$)	Cystic duct (Fig. 2a)	Type 1c	None
Cystic duct ($n = 1$)	Right anterior sectoral duct	Type 1a	None

* The precise sector/segments drained by the aberrant duct could not be defined in three cases, wherein the aberrant duct was incidentally detected at surgery prior to the MRCP era

(i) right hepatic artery crossing anterior to the cyst ($n = 15$),
(ii) replaced right hepatic artery ($n = 8$),
(iii) an accessory right hepatic artery ($n = 3$).

Management of aberrant arterial anatomy

A right hepatic artery crossing anterior to the cyst ($n = 15$): The right hepatic artery was dissected off the

anterior surface of the CHD or the cyst all along its course. The CHD or the cyst was then transected at the appropriate level and the artery transposed posterior to the CHD. The B-EA was reconstructed anterior to the posteriorly transposed right hepatic artery.

A replaced/accessory right hepatic artery: The replaced or accessory right hepatic arteries were carefully dissected off the duct and preserved in 10 of 11 cases. In one case, an accessory right hepatic artery had to be ligated because of avulsion of one of its tributaries while dissecting off the cyst. However, the main right hepatic artery of this child was preserved.

Concomitant aberrant arterial and biliary anatomy

Of the above-mentioned cases, three cases had a concomitant aberrant arterial and biliary ductal anatomy, each of which was managed on its individual merit (Table 1).

Outcome

There was no operative mortality. There was one minor bile leak in the patient who had undergone ductoplasty with a single ostium B-EA, which healed on conservative therapy by postoperative day 5. The duration of long-term follow-up in the aberrant arterial and ductal anatomy group ($n = 40$) ranged from 6 months to 151 months (median: 24 months). Further long-term follow-up as per the protocol defined in "Methods" would continue. The outcome was assessed on the basis of symptoms, liver function tests, ultrasonogram and Tc^{99m}-mebrofenin hepatobiliary scintigraphy as defined in "Methods".

Outcome in aberrant ductal anatomy group ($n = 17$): No patients had symptoms related to biliary or pancreatic pathology. All had normal liver function tests, progressive regression of intrahepatic duct dilatation on ultrasonogram and a satisfactory clearance of radio-isotope from the liver on hepatobiliary scintigraphy as defined in "Methods".

Outcome in aberrant arterial anatomy group ($n = 26$): All patients were asymptomatic with normal liver function tests. In two cases, persistence of intrahepatic duct dilatation, though regressed as compared to preoperative duct diameter, was reported on ultrasonogram at 6 months post-surgery follow-up. On hepatobiliary scintigraphy, these two patients showed a mild delay in clearance of radio-isotope from the liver—total clearance from the liver was achieved at 1.5 h. Since these patients are asymptomatic with normal LFT and the duration of postoperative follow-up is short, they are under close follow-up. The radio-isotope clearance is expected to improve on scintigraphy as the intrahepatic duct dilatation regresses with time. The remaining 24 cases have satisfactory ultrasonogram and hepatobiliary scintigraphy reports at the last follow-up.

Discussion

Variations in the anatomy of the biliary tree are so common that the normal or the conventional anatomy wherein the normal biliary confluence is formed by the union of left and right ducts is present in only 57 % of normal population [3].

Awareness, identification and appropriate management of aberrant biliary anatomy are important because mismanagement of aberrant ducts has serious implications.

The implications of a missed biliary ductal anomaly are: (i) inadvertent transection resulting in a bile leak, which may manifest as a generalized biliary peritonitis or a localized biloma or (ii) an accidental ligation manifesting as pain, cholangitis and segmental atrophy [1–7].

While the relevance of aberrant anatomy has been frequently addressed in the context of live donor liver transplantation and laparoscopic cholecystectomy, there is paucity of literature on the relevance of aberrant ductal anatomy in the management of choledochal cysts, also referred to as congenital choledochal malformations. Todani et al. [13] reported an aberrant posterior duct draining into the distal common hepatic duct in 2 of their 104 cases. The aberrant duct was joined to the hepatic duct at the hilum and a hepaticoduodenostomy was performed with a large stoma created by incisions extending along the lateral walls of both the hepatic ducts and the aberrant ducts.

Narasimhan et al. [14] reported two cases with accessory left-sided hepatic ducts in children with choledochal cyst. None were detected on preoperative imaging, but incidentally detected at surgery. The management was a double ostium anastomosis ($n = 1$) and a ductoplasty with a single ostium anastomosis ($n = 1$), respectively.

Dong et al. reported on two cases of aberrant bile duct in their series of 72 cases. An aberrant segment six duct inserting onto the cystic duct was approximated with the CHD at the confluence followed by a single ostium bilioenteric anastomosis [15].

Major anomalies of the extrahepatic biliary tree have been classified into various types as elucidated in "Methods".

Of relevance to the surgery for congenital choledochal malformations are the following ductal anomalies: (i) the aberrant opening of the right sectoral duct (anterior or posterior) into the CHD or the cyst itself distal to the confluence as was observed in 14 cases in this series; (ii) the ectopic opening of the right sectoral duct into the cystic duct or the cystic duct–CHD junction as observed in two cases in this series (Table 1). In the first scenario, the

aberrant duct, if unrecognized, is at risk of injury during cyst/duct transection. An aberrant duct joining the cystic duct or the cystic duct–common hepatic duct junction is particularly vulnerable to injury in the form of inadvertent ligation or transection while performing cholecystectomy.

In addition, numerous variations related to the joining of the cystic duct with the extrahepatic biliary tree are described [7]. Of significance to the surgery for congenital choledochal malformation is the joining of the cystic duct with the right hepatic duct or the right hepatic sectoral duct as was seen in one of our cases. The cystic duct in this case was significantly dilated with an elongated course running parallel to the right hepatic duct. Here, special care needs to be exercised so as to avoid injury to the right sectoral duct while dissecting off the redundant cystic duct and also carefully repairing the cystic duct–right sectoral duct junction without compromising on the lumen of the right sectoral duct.

Each of the above-mentioned major variations need to be differentiated from a sub-vesical duct, which has been described in 20–50 % of cases [7]. This duct, sometimes deeply embedded in the cystic plate, joins either the common hepatic duct or the right hepatic duct. This duct does not drain any specific liver territory. Although not of major anatomical significance, injury may occur during cholecystectomy if the cystic plate is not preserved and may lead to postoperative biliary leak.

Some authors [5, 6] have also differentiated between a "major" aberrant duct and a "minor" accessory duct, wherein a major aberrant bile duct does not communicate with other biliary segments and is the only bile duct draining a particular segment of the liver. On the other hand, an accessory or "minor" bile duct intercommunicates with other biliary segments and is an additional bile duct draining the same area of the liver. Ligation of an accessory or "minor" bile duct may not cause recurrent cholangitis or focal fibrosis of the liver because of the existence of another draining bile duct, whereas ligation of the aberrant or "major" bile duct results in serious complications. The differentiation between the two can be made on the basis of good preoperative or an intraoperative cholangiogram. All the cases reported in the current series represent a major aberrant duct as documented on the cholangiogram or at surgery.

Prior to the routine use of preoperative MRCP, aberrant ducts were often missed on intraoperative cholangiograms and, hence, incidentally detected during dissection as reported in three cases in this series. An intraoperative cholangiogram missed an aberrant duct either because large volume of contrast was sequestrated within the cyst and, hence, the contrast density or opacification of the proximal biliary tree was unsatisfactory or the aberrant duct was obscured because of overlap by the hugely dilated cyst. A missed aberrant duct because of overlap by dilated extrahepatic biliary tree is an issue specific to the choledochal cyst pathology. Moreover, in the early part of our experience before the year 2005, intraoperative cholangiograms were done without fluoroscopic guidance and the image could be visualized in a single plane/projection only.

However, preoperative MRCP facilitated preoperative detection, delineation of the liver segment or the sector drained by the aberrant duct and its site of insertion on the CHD, the cyst itself or the cystic duct. This facilitated preoperative planning of the surgical strategy. The usefulness of MRCP as a sensitive and non-invasive technique for identifying biliary ductal anomalies in pediatric choledochal cyst has been validated by other authors as well [16]. It is imperative though that the surgeon must make a conscious effort to identify each segmental duct on preoperative cholangiogram. Sometimes, the only clue to an aberrant anatomy may be the inability to visualize one of the sectoral/segmental ducts at or proximal to the confluence, while the aberrant duct itself is obscured because of overlap by a hugely dilated cyst if the MRCP images are viewed and interpreted in limited planes. This was the explanation for missing an aberrant ductal anatomy on preoperative MRCP in three cases in this series. MRCP offers the unique advantage of rotating the image through various angles, thereby eliminating overlap and, hence, the cholangiographic reconstruction should entail rotation through various angles. Sometimes, a small-diameter undilated aberrant duct may show as a faint hair-like structure on an MRCP. Here, a complementary intraoperative cholangiogram (IOC) may define it better, because in an IOC the contrast is injected under some pressure as demonstrated by one case in this series (Fig. 4c).

In addition, a simple technical precaution that would prevent inadvertent injury to an aberrant duct is to initially incise the anterior wall of the cyst significantly distal to the confluence and inspect the cyst from inside for any possible aberrant duct orifice. Circumferential transection may subsequently be done at a more proximal level if deemed necessary.

All major aberrant ducts should be incorporated into the bilio-enteric anastomosis and none should be ligated. The exceptions to this rule would be a sub-vesical duct or a minor accessory duct as described earlier in this study.

The technique of incorporation of the aberrant duct into the bilio-enteric anastomosis would depend on the diameter of the aberrant duct and its proximity with the confluence as elaborated in "Methods". A similar approach has been reported in other case series on the management of aberrant duct anatomy in choledochal cysts [13–15]. However, if a small-diameter aberrant duct is seen to join the CHD or the cyst close to the confluence, the safest approach is to transect the duct distal to the aberrant duct orifice (Fig. 3).

Thus, a thin rim of the CHD or the cyst between the aberrant duct and the confluence is retained and the two are implanted onto the Roux loop with a single ostium anastomosis. This approach was used in 13 of 17 cases in this series. In 8 of these 13 cases, the aberrant duct was inserted onto a relatively normal-caliber CHD proximal to the cystically dilated CBD (Table 1). Hence, this approach did not leave behind residual disease in these eight cases.

Similarly, the implications of a missed arterial anomaly are: (i) injury leading to intraoperative bleed or (ii) accidental ligation which may predispose to delayed postoperative ischemic strictures of hepaticojejunostomy. In the context of post-laparoscopic cholecystectomy bile duct injuries, associated arterial injuries have been reported to adversely affect the outcome of bile duct stricture repair [17]. Moreover, it is important to remember that each artery, whether accessory or replaced, must be preserved because it has its specific terrain of blood supply; each artery is, as a rule, an end artery with no anastomosis inside the liver [18].

The incidence of arterial anomalies has been reported to range from 25 to 75 % [8, 18, 19]. Of relevance to the surgery for congenital choledochal malformation are the following arterial anatomical variations: (i) a replaced or accessory right hepatic artery arising from the superior mesenteric artery: This aberrant right hepatic artery characteristically runs along the free edge of the hepatoduodenal ligament posterolateral to the duct appearing on the medial side of the Calot's triangle and usually running just behind the cystic duct; (ii) a hepatic artery proper crossing anterior to the bile duct. In each of these locations, the anomalous artery may be injured while encircling and transecting the duct.

The hepatic artery proper which is seen to cross anterior to the cyst is dissected along its course and repositioned posterior to the cyst as was done in 15 cases in this series. This was necessary to get adequate space so that the B-EA could be placed at the confluence or carried proximal to the confluence onto the extrahepatic part of the left or right duct as was necessary in five cases with associated congenital stenosis around the hilum.

To summarize, this clinical study emphasizes the need to be aware of, to anticipate and to consciously look for aberrant ductal and arterial anatomy in resection and reconstructive surgery for choledochal cyst. Preoperative MRCP facilitated preoperative detection and delineation of the precise anatomy of the aberrant duct and, hence, planning out the surgical strategy. However, aberrant ducts may occasionally be missed on MRCP as well because of overlap by a hugely dilated cyst, especially because undilated aberrant ducts may be visualized faintly. Hence, MRCP reconstruction should necessarily entail rotating the images through various angles to eliminate overlap by the dilated biliary tree. All major aberrant ducts should be incorporated into the biliary-enteric anastomosis and not ligated; the technique of incorporation into the biliary-enteric anastomosis would be governed by the distance of the aberrant duct from the confluence and the diameter of the duct. Likewise, one must palpate and consciously look for arterial pulsations anterior and postero-lateral to the duct before encircling and duct transection. All aberrant arteries, whether accessory or replaced, should be carefully preserved.

Acknowledgments Dr. Basant Kumar and Dr. Vijay Upadhyaya assisted in performing the surgical procedure in cases operated since 2009. Dr. Zafar Neyaz gave useful inputs about the technical details of MRCP. Dr. S Gambhir gave useful inputs about the technical details of hepatobiliary scintigraphy. Mr. Anil Kumar prepared the line diagrams.

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Ohkuba M, Nagino M, Kamiya J, Yuasa N, Oda K, Arai T, Nishio H, Nimura Y (2004) Surgical anatomy of the bile ducts at the hepatic hilum as applied to live donor liver transplantation. Ann Surg 239:82–86
2. Marcos A, Ham JM, Fisher RA, Olzinski AT, Posner MP (2000) Surgical management of anatomical variation of right lobe in living donor liver transplantation. Ann Surg 231:824–831
3. Deshpande RR, Heaton ND, Rela M (2002) Surgical anatomy of segmental liver transplantation. BJS 89:1078–1088
4. Larobina M, Nottle PD (2005) Extrahepatic biliary anatomy at laparoscopic cholecystectomy: is aberrant anatomy important? ANZ J Surg 75:392–395
5. Hirao K, Miyazaki A, Fujimoto T, Isomoto I, Hiyashi K (2000) Evaluation of aberrant bile ducts before laparoscopic cholecystectomy: helical CT cholangiography versus MR cholangiography. AJR 175:713–720
6. Izuishi K, Toyama Y, Nakano S, Goda F, Usuki H, Masaki T, Macta H (2005) Preoperative assessment of the aberrant bile duct using multislice computed tomography cholangiography. Am J Surg 189:53–55
7. Blumgart LH, Hann LE (2000) Surgical and radiological anatomy of the liver and biliary tree. In: Blumgart LH, Fong Y (eds) Surgery of the liver and biliary tract, vol 1, 3rd edn. W. B. Saunders Company Ltd, Edinburgh, pp 13–34
8. Hiatt JR, Gabbay J, Busuttil RW (1994) Surgical anatomy of the hepatic arteries in 1,000 cases. Ann Surg 220:50–52
9. Hill R, Parsons C, Farrant P, Sellars M, Davenport M (2011) Intrahepatic duct dilatation in type 4 choledochal malformation: pressure related, postoperative resolution. J Pediatr Surg 46: 299–303
10. Sarkari A, Gambhir S, Kumar A, Saxena R, Kapoor VK, Sikora SS (2004) Evaluation of bilioenteric anastomosis using quantitative hepatobiliary scintigraphy. Hepatogastroenterology 51: 1267–1270
11. Todani T, Watanabe Y, Toki A, Morotomi Y (2003) Classification of congenital biliary cystic disease: special reference to type Ic and type IVA cysts with primary ductal stricture. J Hepatobiliary Pancreat Surg 10:340–344

12. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K et al (1977) Congenital bile duct cyst: its classification, operative procedures and review of 37 cases including cancer arising from choledochal cyst. Am J Surg 134:263–269
13. Todani T, Watanabe Y, Toki A, Ogura K, Wang Z-Q (1998) Co-existing biliary anomalies and anatomical variants in choledochal cysts. BJS 85:760–763
14. Narasimhan KL, Chowdhary SK, Rao KLN (2001) Management of accessory hepatic ducts in choledochal cysts. J Pediatr Surg 36:1092–1093
15. Dong Q, Jiang B, Zhang H, Jiang Z, Lu H, Yang C, Cheng Y, Hao X (2006) Management strategy for congenital choledochal cyst with co-existing intrahepatic dilatation and aberrant bile duct as well as other complicated biliary anomalies. Yonsei Med J 47:826–832
16. Saito T, Hishiki T, Terui K, Sato Y, Mitsunaga T, Terui E, Nakata M, Takenouchi A, Matsuura G, Yahata E, Ohno S, Sato H, Yanagawa N, Masuda Y, Yoshida H (2011) Use of preoperative, 3-dimensional magnetic cholangiopancreaticography in pediatric choledochal cysts. Surgery 149:569–575
17. Schmidt SC, Settmacher U, Langrehr JM, Neuhaus P (2004) Management and outcome of patients with combined bile duct and hepatic arterial injuries after laparoscopic cholecystectomy. Surgery 135(6):613–618
18. Michels NA (1962) Newer anatomy of the liver and its variant blood supply and collateral circulation. Am J Surg 112:337–347
19. Nelson TM, Pollak R, Jonasson O, Abcarian H (1988) Anatomic variants of the coeliac, superior mesenteric and inferior mesenteric arteries and their clinical relevance. Clin Anat 1:75–91