ECRONICON OPEN ACCESS

EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Series

Congenital Bile Duct Cysts in Adults: Management and Outcome of Clinical Cases

B Bouibaouen*, M Salihoun, M Acharki and N Kabbaj

Functional Endoscopy Department, Ibn Sina Hospital, Rabat, Morocco

***Corresponding Author:** B Bouibaouen, Functional Endoscopy Department, Ibn Sina Hospital, Rabat, Morocco.

Received: September 06, 2021; **Published:** October 29, 2021

Abstract

Introduction: Congenital cystic dilation of the common bile duct (CDCBD) is a congenital malformation, represented by a segmental ectasia with variable types and dimensions. It's generally limited to the common bile duct, but an association of the hepatic ducts dilatation can be observed.

Results: We report three clinical cases of congenital cystic dilatation of the common bile duct, over a period of May 2015 to April 2018. The average age is 70 years, with extreme ages of 55 and 85 years. The sex ratio is 1/3 with a female predominance. 1 patient presented an abdominal pain, 1 was revealed by a complication and the diagnosis was accidentally discovered. 1 of our patients presented a cytolysis due to a viral hepatitis. Ultrasound was performed in all 3 patients, and in all 3 cases it demonstrated dilation of the CBD without any obstacle. And bili MRI confirmed the diagnosis. Our patients have not been operated.

Discussion: The classic symptomatic triad of CDCBD is abdominal pain, jaundice, and a palpable abdominal mass. However, the CDCBD can be accidentally discovered. Biological cholestasis and hepatic cytolysis can be found. MRI is currently the reference examination in the diagnosis of CDCBD. Todani's Type I dilation remains by far the most frequent case of CDCBD, followed by type Iva. Main complications of the CDCBD are infections, lithiasis, secondary biliary cirrhosis or malignant degeneration. Endoscopic treatment is done by an endoscopic sphincterotomy to ensure effective biliary drainage. Surgery is the treatment of choice for congenital cystic dilation of the main bile duct. The excision will prevent degeneration of the cyst wall and bile ducts. Percutaneous radiological drainage is indicated in palliative situations.

Conclusion: In patients who have not received surgical management, regular clinical, biological and morphological monitoring is necessary in order to detect the occurrence of any complication at an early stage. However, to date, there are no recommendations on the monitoring of these unoperated patients age of the patients and their medical background is important to analyze, to make the assure the best medical care.

Keywords: *Congenital Cystic Dilatation; Common Bile Duct; CDCBD*

Introduction

Congenital cystic dilation of the common bile duct (CDCBD) is a congenital malformation, represented by a segmental ectasia with variable types and dimensions. It's generally limited to the common bile duct, but an association of the hepatic ducts dilatation can be observed [1,2]. The dilation can be total or segmental. According to Todani, they are grouped into five types based on the location, extent and type of bile duct dilation which can be either aneurysmal or spindle-shaped or rarely diverticular [3].

We report three clinical cases of cystic dilatation of the common bile duct, which will be discussed in the light of the data in the literature, and whose diagnosis was suspect on endoscopic ultra sonography and confirmed on bili MRI or on cholangiography.

Case 1

F.A, 85 years old, with a history of hypertension.

The patient was initially seen on the month of April 2014 for management of chronic hepatitis C, genotype 1b with a viral load of 6.22 log/ml.

Clinical examination found a patient with no fever, anicteric, a BMI of 24 kg/m^2, the abdominal examination was normal. The biological assessment demonstrated cytolysis with TGO at 133 IU/l (4xN) and TGP at 125 IU/l (2.5xN), without biological cholestasis: total bilirubin at 9.2 mg/l, PAL at 71 IU/l, GGT at 27 IU/l. The patient did not have co-infection with hepatitis B virus (HBsAg, anti HBsAb, and anti HBcAb: negative).

Abdominal ultrasound revealed cirrhosis without focal lesions, neither signs of PHN nor ascites, an alithiasis gallbladder and a 10 mm dilated CBD without echo-visible obstruction. Echo-endoscopy showed dilated CBD in its intrahepatic portion to 14.2 mm with cystic dilation without visible obstacle associated with a slight dilation of the left hepatic ducts, very suggestive of choledochal cystic dilatation (CCD). Bili-MRI confirmed the diagnosis of CCD type Ic.

A follow-up in consultation with periodic MRI (every 6 months) was provided in the context of his cirrhosis.

The patient died of his cirrhosis' complications.

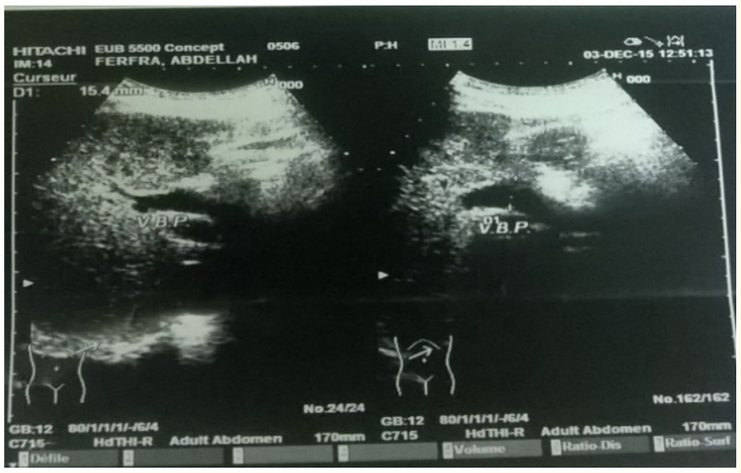

Figure 1: *US showing CBD dilatation (Case 1).*

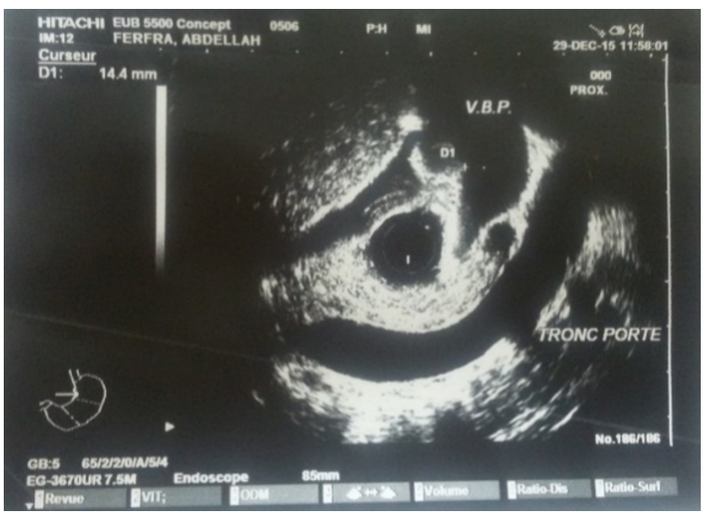

Figure 2: *EUS showing a CDCBD (Case 1).*

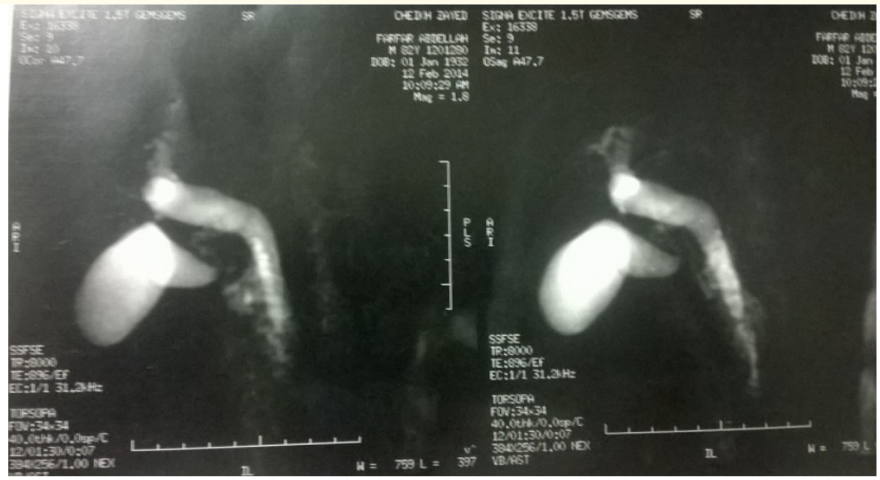

Figure 3: *Bili MRI showing a CDCBD type Ic of Todani (Case 1).*

Case 2

D.S, 70 years old, with no pathological history, who presented with isolated right hypochondrium pain without any other symptoms.

The clinical examination found an anicteric, afebrile patient with a BMI of 20 kg/m^2, the abdominal examination was normal. Liver function tests did not reveal cytolysis or cholestasis. Abdominal ultrasound showed a hydatid cyst of the liver and a CBD dilation without visible obstruction or gallstones.

Abdominal CT showed hepatic hydatid cyst and dilation of the CBD.

Endoscopic ultrasound revealed cystic dilation of the proximal portion of the CBD (19.4 mm) without visible obstruction. In its distal portion, the CBD is thin (5.5 mm) with no obstacle image. Bili-MRI confirmed CCD (18 mm) type Ib, with a normal gallbladder.

Surgery was proposed for the hydatic liver cyst. The CCD was respected due to the patient's advanced age.

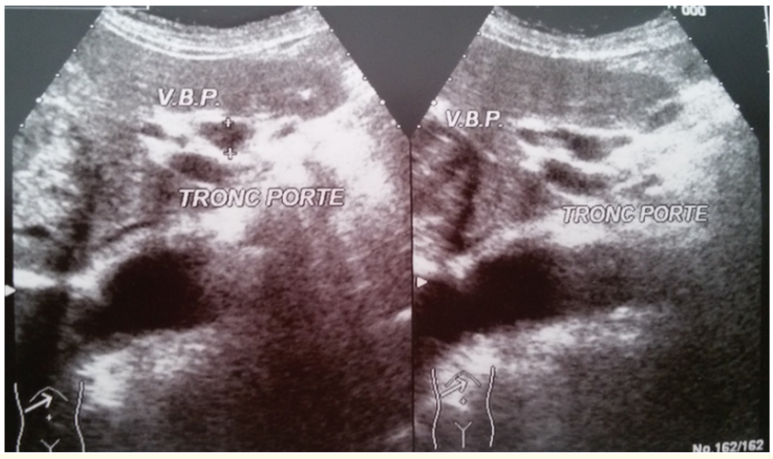

Figure 4: *US showing a CBD dilatation (Case 2).*

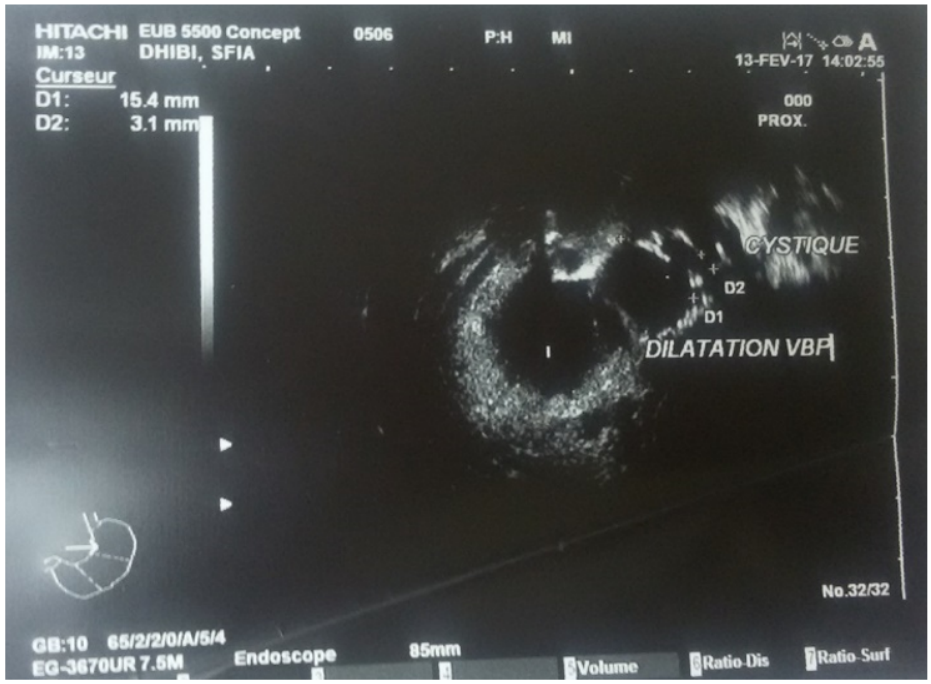

Figure 5: *EUS showing a CDCBD (Case 2).*

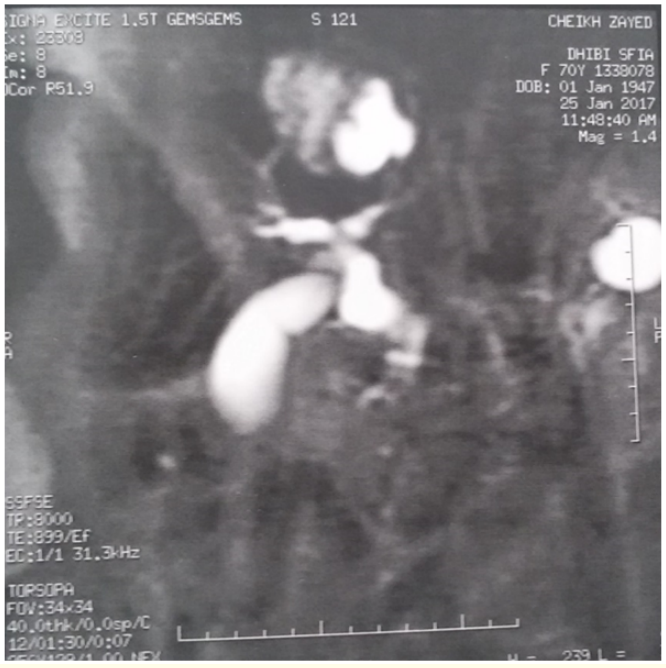

Figure 6: *Bili-MRI showing a CDCBD type Ib of Todani (Case 2).*

Case 3

L.A, 55 years old, with no pathological history.

In 2015, she presents the first episode of acute pancreatitis. After a free interval of 20 days, the patient was rehospitalised for recurrent acute pancreatitis. The clinical course was favorable and the patient was lost to follow-up until October 2017: where she was rehospitalized for a 3rd episode of acute pancreatitis, Balthazar E. Liver function tests were normal.

Abdominal ultrasound showed dilation of the CBD to 14 mm with dilation of the intrahepatic bile ducts without visible obstruction. Bili-MRI concluded on a CBD dilatation (16 mm) without any clearly visible obstruction.

ERCP showed a cystic dilation of the terminal part of the common bile duct (20 mm) and hepatic bile ducts with a low cystic duct inserted and intracystic lithiasis that have been extracted.

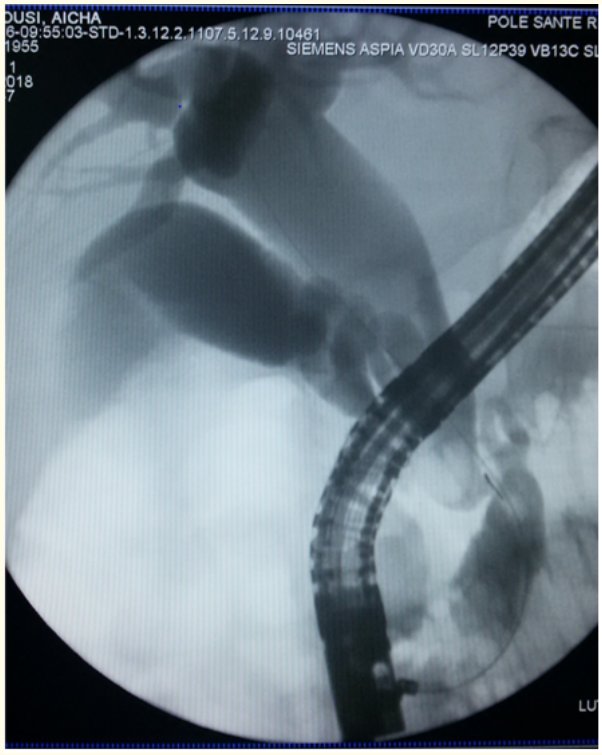

Figure 7: *ERCP showing a CDCBD type IVa.*

Discussion

CDCBD is a rare anatomo-clinical entity with an incidence ranging: 1/13,500 births in the United States, 1/15,000 births in Australia [4,5]. The rate is remarkably higher in Asian populations with a reported incidence of 1/1000 births, and about two-thirds of cases occur in Japan [6,7]. CDCBD ranks second after bile duct atresia [8].

CDCBD is found mainly in children, adolescents and young adults with 80% of cases being observed before the age of 10 years [6,7].

Series	Country	Cases	Duration of the study (years)	Middle age (years)	Age extremes (years)	Sex ratio M/F
Atkinson., *et al*. 2003 [10]	Ecosse	16	8	23	18-49	1/3
Colin., *et al*. 2006 [12]	Singapore	32	14	41	18-74	1/ 3,5
Mannai., *et al*. 2006 [11]	Tunisie	18	20	53,1	5-80	1/2
Cho MJ., *et al*. 2011 [2]	Japon	204	14	40	20-60	1/3
Lei Gong., *et al*. 2012 [15]	USA	221	9	37	13-77	1/3,5
Norman., *et al*. 2015 [9]	Oman	10	15	31	16-38	1/4

Table 1: *Epidemiological data of CDCBD.*

The congenital nature is underlined by the notion of a case discovered on antenatal ultrasound. The adult forms correspond to cases that exist from childhood and remain undiagnosed because they are well tolerated [9].

The female predominance is described by all the authors with a sex ratio of 3.5 to 4. This female predominance can reach 77% to 80% in some studies [9,10].

In our series, the diagnosis of CDCBD was made in 3 patients over a period of May 2015 to April 2018. The average age is 70 years, with extreme ages of 55 and 85 years. The sex ratio is 1/3 with a female predominance.

Many theories have been proposed to explain the origin of these malformations, the best known of which are: the mechanical theory, which shows that the common bile duct dilatation is a result of the lower bile duct stenosis [16] and the congenital theory which admits as a mechanism a defective embryological development of the bile ducts according to the Yotsuyanagi hypothesis [17] and an anomaly of the biliopancreatic junction according to the Babitt hypothesis. This theory remains the most commonly accepted [18,19].

A genetic Chinese study demonstrated the heterogeneity of the genetic profile in patients with CDCBD and the concomitant presence of several mutations in several genes, which may explain the rarity of this pathology [20].

The cystic wall is formed of a sclerosis connective tissue, within a few rare elastic fibers and muscle cells lined with a squamous biliary-type epithelium which is almost destroyed, reduced to a few areas of flattened or cubic cells, inflammatory, ulcerated, and covered with a fibrinoleukocytic coating, resting on a basal conjunctive condensation [21,22]. Anatomists insist on neuro-ganglionic agenesis at the cystic wall, which is believed to be the primary causal lesion. Degeneration into adenocarcinoma is possible and has been observed mainly in Japan [4,23].

The classic symptomatic triad of CDCBD is abdominal pain, jaundice, and a palpable abdominal mass [24]. However, this triad is only present in less than a third of cases, in almost two thirds of cases patients have one or two of the three symptoms [15,23,25,26]. Also, patients can remain asymptomatic, and the CDCBD is accidentally discovered [9,12]

Series	Cases	Pain	Jaundice	Abdominal mass	Symptomatic triad
Atkinson., *et al*. 2003 [10]	16	16 case (100%)	0 case (0%)	0 case (0%)	0 case (0%)
Mannai., *et al.* 2006 [11]	18	18 case (100%)	10 case (55%)	0 case (0%)	0 case (0%)
Colin., *et al.* 2006 [12]	32	29 case (91%)	13 case (41%)	8 case (25%)	4 case (13%)
Lei Gong., *et al*. 2012 [15]	221	177 case (80%)	52 case (24%)	N.P	N.P
Norman., *et al.* 2015 [9]	10	10 case (100%)	7 case (70%)	2 case (20%)	2 case (20%)

Table 2*: Reported symptomatology.*

Biological cholestasis and hepatic cytolysis are found in 50% to 69% of the cases [10]. Recent publications report the association of CDCBD with biliopancreatic junction anomaly in approximately 90 to 100% of cases [27] and an elevated concentration of pancreatic enzymes in the bile. When pressure increases in the common bile duct by obstructive cholangitis, pancreatic bile enzymes can pass into the blood stream. The increase in lipasemia is also observed in acute pancreatitis, secondary to CDCBD [28].

Ultrasound can contribute to the initial diagnosis of CDCBD in 71 to 97% of cases [29,30].

On ultrasound, the cyst appears as an oval mass, easily dissociated from the gallbladder, with sharp boundaries, anechoic with posterior reinforcement and fluid-like appearance which may contain stones. A communication between the CBD and this cyst is essential to confirm the diagnosis, but the volume of dilation can lead to confusion with the gallbladder [24].

Ultrasound was performed in all 3 patients, and in all 3 cases it demonstrated dilation of the CBD without any obstacle.

MRI is currently the reference examination, allowing a multi-plane exploration of the bilio-pancreatic pathways, in particular the diagnosis of CDCBD [15,31,32]. Bili MRI not only allows the positive diagnosis of CDCBD in 96% of cases, but also the precision of the classification of this dilation (Table 3: classification of Todani) with a specificity exceeding 90%, a sensitivity exceeding 80%, a positive predictive value exceeding 80% and a negative predictive value approaching 95%, thus becoming the best radiological examination of the CDCBD [32].

Type I:
Ia: Sacciform dilatation
Ib: Segmental dilatation
Ic: Fusiform dilatation
Id: Multiple extra hepatic dilatation
Type II: CBD diverticulum
Type III: Choledocele
Type IV:
IVa: IH and CBD sacciform dilatation
IVb: IH and CBD segmental dilatation
IVc: IH and CBD fusiform dilatation
V: Caroli disease
VI: Cystic duct dilatation

Table 3: *Todani classification [37].*

Bili-MRI was performed in our 3 patients, confirmed the diagnosis and staged the CDCBD in 2 cases. However, in the third case, Bili-MRI showed a CBD dilatation without visible obstacle, without confirmation of the diagnosis of CDCBD.

Ultrasound endoscopy is a technique combining both endoscopy and ultrasound.

The advantage of this technique is that it makes it possible to study the bile ducts and to overcome the fatty and gas interpositions that often prevent proper visualization of the biliary structures during traditional ultrasound [33].

Ultrasound endoscopy contributes to the diagnosis of biliary malformations, in particular CDCBD [34,35], but also the visualization of bile duct stones.

in 2 of our patients, EUS showed a dilation of the CBD without visualization of an obstacle.

The ERCP shows the precise anatomy of the malformation (type, size, caliber), the supra-cystic and intrahepatic bile ducts, the accessory bile duct, the sub-cystic common bile duct and the biliopancreatic junction [36]. The incidence of acute post-ERCP pancreatitis is higher in patients with CDCBD [30]. Thus, this technique is indicated essentially for therapeutic purposes.

ERCP was performed in a 1 patient for diagnostic and therapeutic purposes. In fact, it initially demonstrated CDCBD during the opacification of the bile ducts and to extract intra-cystic lithiasis debris in a second step.

According to Todani's classification, 1 of our patients had a type Ic CDCBD, one patient a type Ib and the third patient had a type IVa CDCBD. Analysis of the different case studies described in the literature shows that Todani's Type I dilation remains by far the most frequent case of CDCBD, followed by type IVa [9-12].

Series	Cases	Type I	Type II	Type III	Type IVa	Type IVb	Type V
Atkinson., *et al*. 2003 [10]	16	13 case (81%)	0 case	0 case	3 case (19%)	0 case	0 case
Coli., *et al*. 2006 [12]	32	27 case (85%)	2 case (6%)	0 case	2 case (6%)	0 case	1 case (3%)
Mannai., *et al*. 2006 [11]	18	11 case (61%)	0 case	0 case	0 case	0 case	7 case (39%)
Cho MJ., *et al*. 2011 [2]	204	116 case (57%)	1 case (0,5%)	0 case	86 case (42%)	0 case	1 case (0,5%)
Lei Gong., *et al*. 2012 [15]	221	168 case (76%)	3 case (1,3%)	3 case (1,3%)	26 case (11,7%)		21 case (9,5%)
Norman., *et al* 2015[9]	10	8 case (80%)	1 case (10%)	1 case (10%)	0 case	0 case	0 case

Table 4*: Todani's classification in different published studies.*

Patients are mono or bi-symptomatic for a long time in the classic form and sometimes even asymptomatic, and the CDCBD only manifests itself through complications [24,38]: infections, lithiasis, secondary biliary cirrhosis or malignant degeneration.

Chronic inflammation and activation of bile salts would then lead to the high frequency of lithiasis and malignant transformation. This mutagenic activity may be linked to the K-ras oncogene and to the mutation in the p53 gene [39]. In addition, significant concentrations of carcinogenic substances have been identified in the bile contents in the presence of biliopancreatic junction anomaly [21]. Radiological diagnosis of cyst degeneration is based on the search for localized or diffuse thickening of the cystic wall.

Endoscopic treatment is done by an endoscopic sphincterotomy to ensure effective biliary drainage [40]. It also allows spontaneous or instrumental evacuation of gallstones.

Surgery is the treatment of choice for congenital cystic dilation of the main bile duct. The excision will prevent degeneration of the cyst wall and bile ducts. Cysto-digestive derivations are to be avoided [41,42].

Percutaneous radiological drainage is indicated in palliative situations, but also after failure of endoscopic and surgical treatment [43].

One of our patients underwent an ERCP with intracystic calculus extraction and was referred to another center for further treatment and was not reviewed in consultation. In the other two cases, the surgery was not performed because of the advanced age of patients: one is still followed in consultation and has no complications, the second died of his cirrhosis' complications.

In patients who have not received surgical management, regular clinical, biological and morphological monitoring is necessary in order to detect the occurrence of any complication at an early stage. However, to date, there are no recommendations on the monitoring of these unoperated patients.

Conclusion

Cystic dilatation of common bile duct is a congenital disease. More observed in Asian countries, female and young patients.

The main used classification is Todani classification. Type I is the most frequent type.

CDCBD can be discovered in asymptomatic patients or revealed by its complications.

MRI is important for certain diagnosis and classification precision. ERCP is used for therapeutic purpose

Best treatment is surgical excision of the cystic dilatation, which prevent the malign degeneration. However, age of the patients and their medical background is important to analyze, to make the assure the best medical care.

Bibliography

1. Makin E and Davenport M. "Understanding choledochal malformation". *Archives of Disease in Childhood* 97.1 (2012): 69-72.
2. Cho MJ., *et al*. "Surgical experience of 204 cases of adult choledochal cyst disease over 14 years". *World Journal of Surgery* 35.5 (2011): 1094-1102.
3. Kianmanesh R., *et al*. "Pancreato-biliary maljunctions and congenital cystic dilatation of the bile ducts in adults". *Journal de Chirurgie* 138.4 (2001): 196-204.
4. Howard ER. "Surgery of liver disease in children". Butterworth-Heinemann Medical (1991).
5. Gigot JF., *et al*. "Bile duct cysts: a changing spectrum of presentation". *Journal of Hepato-Biliary-Pancreatic Surgery* 3.4 (1996): 405-411.
6. Ahluwalia A., *et al*. "Choledochal Cyst-A Rare Entity". *Indian Journal of Radiology and Imaging* 12.4 (2002): 491.
7. Bricha M and Dafiri R. "Une cause inhabituelle d'un abdomen aigu chez l'enfant: la rupture spontanée d'un kyste du cholédoque". *Journal de Radiologie* 88.5 (2007): 692-693.
8. Todani T., *et al*. "Carcinoma related to choledochal cysts with internal drainage operations". *Surgery, Gynecology and Obstetrics* 164.1 (1987): 61-64.
9. Machado NO., *et al*. "Choledochal Cyst in Adults: Etiopathogenesis, Presentation, Management, and Outcome—Case Series and Review". *Gastroenterology Research and Practice* (2015).
10. Atkinson HDE., *et al*. "Choledochal cysts in adults and their complications". *Hpb* 5.2 (2003): 105-110.
11. Mannai S., *et al*. "Congenital cystic dilatation of bile ducts". *In Annales de Chirurgie* 131.6-7 (2006): 369-374.

12. Woon CYL., *et al*. "Adult choledochal cysts: an audit of surgical management". *ANZ Journal of Surgery* 76.11 (2006): 981-986.

13. Cussenot O., *et al*. "Congenital dilatation of the common bile duct". *Chirurgie Pediatrique* 28.1 (1987): 8-19.

14. Frank JL., *et al*. "Antenatal observation of a choledochal cyst by sonography". *American Journal of Roentgenology* 137.1 (1981): 166-168.

15. Gong L., *et al*. "Clinical analysis of 221 cases of adult choledochal cysts". *The American Surgeon* 78.4 (2012): 414-418.

16. Desmet VJ. "Congenital diseases of intrahepatic bile ducts: variations on the theme "ductal plate malformation". *Hepatology* 16.4 (1992): 1069-1083.

17. Yamaguchi M. "Congenital choledochal cyst: analysis of 1,433 patients in the Japanese literature". *The American Journal of Surgery* 140.5 (1980): 653-657.

18. Le Roy B., *et al*. "Pancreaticobiliary maljunction and choledochal cysts: from embryogenesis to therapeutics aspects". *Surgical and Radiologic Anatomy* 38.9 (2016): 1053-1060.

19. Babbit DP. "Congenital choledochal cyst: new etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb". *Annals of Clinical Radiology* 12 (1969): 231-240.

20. Wong JK., *et al*. "Genetic study of congenital bile-duct dilatation identifies de novo and inherited variants in functionally related genes". *BMC Medical Genomics* 9.1 (2016): 1-10.

21. Funabiki T., *et al*. "Biliary carcinogenesis in pancreaticobiliary maljunction". *Journal of Hepato-Biliary-Pancreatic Surgery* 4.4 (1997): 405-411.

22. Ronat R. "Pseudo-Kyste De Hepatocholedoque Associe A Une Dilatation Segmentaire De La Portion Proximale Des Voies Biliaires Intrahepatiques (1979).

23. Singham J., *et al*. "Choledochal cysts: part 1 of 3: classification and pathogenesis". *Canadian Journal of Surgery* 52.5 (2009): 434.

24. C Singham J., *et al*. "Choledochal cysts: part 2 of 3: Diagnosis". *Canadian Journal of Surgery* 52.6 (2009): 506.

25. Shi LB., *et al*. "Diagnosis and treatment of congenital choledochal cyst: 20 years' experience in China". *World Journal of Gastroenterology* 7.5 (2001): 732.

26. Erraoui MG. "Dilatation kystique congénitale du cholédoque (A propos de 09 cas) (1990).

27. Visser BC., *et al*. "Congenital choledochal cysts in adults". *Archives of Surgery* 139.8 (2004): 855-862.

28. Kabbaj N., *et al*. "Apropos of a case of acute pancreatitis revealing cystic dilatation of the common bile duct". *Journal De Radiologie* 79.11 (1998): 1393-1397.

29. Akhan O., *et al*. "Choledochal cysts: ultrasonographic findings and correlation with other imaging modalities". *Abdominal Imaging* 19.3 (1994): 243-247.

30. Sugiyama M., *et al*. "Anomalous pancreaticobiliary junction shown on multidetector CT". *American Journal of Roentgenology* 180.1 (2003): 173-175.

31. Irie H., *et al*. "Value of MR cholangiopancreatography in evaluating choledochal cysts". *AJR. American Journal of Roentgenology* 171.5 (1998): 1381-1385.

32. Park DH., *et al*. "Can MRCP replace the diagnostic role of ERCP for patients with choledochal cysts?" *Gastrointestinal Endoscopy* 62.3 (2005): 360-366.

33. O Croizet., *et al*. "Méthodes d'exploration des voies biliaires". *Encyclopédie Médico Chirurgicales Hépatologie* (1998): 11.

34. Sugiyama M and Atomi Y. "Endoscopic ultrasonography for diagnosing anomalous pancreaticobiliary junction". *Gastrointestinal Endoscopy* 45.3 (1997): 261-267.

35. Yusuf TE and Bhutani MS. "Role of endoscopic ultrasonography in diseases of the extrahepatic biliary system". *Journal of Gastroenterology and Hepatology* 19.3 (2004): 243-250.

36. Blickman JG., *et al*. "Pediatric radiology: the requisites e-book". *Elsevier Health Sciences* (2009).

37. Todani T., *et al*. "Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst". *The American Journal of Surgery* 134.2 (1977): 263-269.

38. Shi LB., *et al*. "Diagnosis and treatment of congenital choledochal cyst: 20 years′ experience in China". *World Journal of Gastroenterology* 7.5 (2001): 732.

39. Fieber SS and Nance FC. "Choledochal cyst and neoplasm: a comprehensive review of 106 cases and presentation of two original cases". *The American Surgeon* 63.11 (1997): 982-987.

40. Shah SK., *et al*. "Therapeutic biliary endoscopy". *Endoscopy* 34.01 (2002): 43-53.

41. Todani T., *et al*. "Choledochal cyst, pancreatobiliary malunion, and cancer". *Journal of Hepato-Biliary-Pancreatic Surgery* 1.3 (1994): 247-251.

42. Chijiiwa K and Koga A. "Surgical management and long-term follow-up of patients with choledochal cysts". *The American Journal of Surgery* 165.2 (1993): 238-242.

43. Farhat LB., *et al*. "Drainage biliaire percutané: efficacité et complications à court et à moyen terme à travers une série de 50 cas". *Tunisie Médicale* 87.1 (2009): 50-54.

Volume 8 Issue 11 November 2021