

Contents lists available at [ScienceDirect](http://www.sciencedirect.com)

## International Journal of Surgery

journal homepage: [www.journal-surgery.net](http://www.journal-surgery.net)

## Review

## Oriental cholangiohepatitis – Is our surgery appropriate?



Fazl Q. Parray\*, Mehmood A. Wani, Nazir A. Wani

Department of General Surgery, Sher I Kashmir Institute of Medical Sciences, Srinagar, Kashmir, 190011, India

## HIGHLIGHTS

- How to grade the disease before surgery.
- Use of access loops internal and external.
- Proper referral to high volume centers.
- Avoiding unwanted surgeries.

## ARTICLE INFO

## Article history:

Received 26 April 2014

Accepted 29 May 2014

Available online 8 June 2014

## Keywords:

Oriental  
Cholangiohepatitis  
Lithogenesis  
Cholangitis  
Access loops

## ABSTRACT

Oriental cholangiohepatitis, or recurrent pyogenic cholangitis is only noted in certain parts of the world, especially South East Asia. Due to increasing immigration the disease is now being seen in western countries also. Treating physicians may face difficulty in managing such cases due to lack of exposure. Furthermore management of such cases is not standardized because of lack of a universally accepted classification system. Here we review the disease and share our long experience with management of these patients.

© 2014 Surgical Associates Ltd. Published by Elsevier Ltd. All rights reserved.

## 1. Introduction

Oriental cholangiohepatitis (OCH), an affliction of poverty and rural environment, is characterized by intrahepatic pigment stones as the pathophysiological end result of the condition. OCH is endemic to all countries rimming South China Sea. Improving life standards account for the decreasing trend of OCH in developing countries while increasing immigrant population partly accounts for the rise noticed in developed countries. Etiology of the condition has been theorized variously but nothing concrete has been established so far. Clinically it is characterized by repeated attacks of abdominal pain, fever and jaundice caused by intrahepatic ductal calculi and strictures [1]. Need for multidisciplinary approach i.e. integrating interventional radiology, interventional endoscopy and surgery is agreed world-over but established management protocols for the condition are yet to surface [2]. In view of its occurrence as an evident morbid disease in socioeconomically

productive age group, economic impact of the condition is immense.

## 2. Incidence

Recurrent pyogenic cholangitis is endemic to South East Asia, where it was first described [3]. In developed countries, though the disease is rare but it has an increasing trend as is seen in United Kingdom and United State attributed to increase in immigrant population [4,5]. The decreasing trend in South East Asia has been historically attributed to the westernization of developing countries [6]. However, Park et al. has reported that despite increasing westernization of Korean life style, the incidence of hepatolithiasis has remained the same [7]. RPC in India is seen exclusively in Kashmir as reported by Khuroo et al. [8]

## 3. Etiopathogenesis

In hepatolithiasis bacterial infection via biliary tract is thought to be the most important step in lithogenesis [9,10]. *Escherichia coli* (*E. Coli*), *Klebsiella*, *Streptococcus* and *Pseudomonas* are the most frequent isolates from the bile of such patients [9]. ®-Glucuronidase

\* Corresponding author. 44-Rawal Pora, Govt Housing Colony, Sanat Nagar, Srinagar, 190005, J&K, India.

E-mail addresses: [fazlparray@rediffmail.com](mailto:fazlparray@rediffmail.com), [fazlparray@gmail.com](mailto:fazlparray@gmail.com) (F.Q. Parray).

from gram negative bacteria, particularly *E. coli* may be involved in deconjugation of bilirubin, a process followed by the formation of calcium-bilirubinate, a major component of brown pigment stones. Zen Y et al., proposed the possible role of lipopolysaccharide (LP), a component of gram negative bacteria in aberrant mucin expression by biliary tree epithelial cells (increased expression of gel-forming apomucin viz MUC2 and MUC5AC), considered to be important in the lithogenesis of hepatolithiasis [11].

In addition low protein diet may result in a relative deficiency of glucoro l-4 lactone in the bile [1], with decreased bactericidal and  $\beta$ -glucuronidase inhibition properties thereby favoring deposition of pigment stones in the biliary tree [12–14]. Diet low in saturated fats is also believed to promote bile stasis and so biliary lithogenesis as it is thought to lack the stimulant for the release of cholecystokinin. As majority of the cases of recurrent pyogenic cholangitis (RPC) occur at a young age some authorities believe that congenital anomalies of the ductal system predispose bile stasis subsequently resulting in stone formation [15]. Presence of lymphopenia and reduced CD4 counts in some patients with OCH suggests the role of some form of immune depression [16]. The suggestion of multi-factorial etiology, with complex interaction of factors like biliary parasites, enteric pathogens and malnutrition, is the most plausible explanation for etiopathogenesis in literature at present.

#### 4. Pathology

Probably the dilatation of intra and extrahepatic biliary ducts with luminal pigment stones, sludge and parasitic, bacterial and cellular debris constitute the pathological hall mark of OCH. Ductal wall fibrosis most markedly involves the central major lobar and segmental ducts. Smaller peripheral ducts appear sharply truncated and cutoff giving the overall picture of “Pruned Tree”. The left hepatic duct particularly left lateral segmental duct is affected more severely and is involved early in the course of the disease. Left biliary tree particularly at confluences is more vulnerable to strictureous ductal fibrosis [12].

#### 5. Management

Difficult access to the parts of biliary tree to which the disease is commonly distributed and failure to address the yet unknown basic etiopathogenetic mechanism, adds-up to the quagmire which the condition poses to the clinician. Integrated use of interventional endoscopy, interventional radiology and surgery, skillfully tailored to the thoroughly evaluated individual patient holds the key for attaining optimal results. A combination of all treatment modalities, on selective basis has been advocated and by using a systematic approach improvement in treatment results has been reported for recurrent pyogenic cholangitis [17].

Rate of complete stone clearance by hepatic resection and PTCSL achieved in such patients has been as high as 84–100% and 72.9–92% respectively [18–24]. Stone recurrence has been reported between 9.5 and 16% of hepatic resection and 32.6–40% with cholangioscopic lithotomy [23,24]. Complication rates have been reported to be approximately 12–40% with hepatic resection and 14.5–22% with Percutaneous transhepatic cholangioscopic lithotomy (PTCSL) [19,22,23,25]. Wound infection, subphrenic abscess, bile leakage, cholangitis and sepsis were reported with hepatic resection, and hemobilia, extreme pain, nausea or vomiting, cholangitis and sepsis with PTCSL. Hemobilia is a serious complication that can be fatal [18,19,22,23,25,26]. The mortality rates have been reported at 1.7–3.2% with hepatic resections and 2.1–8% with PTCSL [19,23,25,27].

Strategies for patients with bilateral involvement remain controversial. Hepatic resections are still considered on more diseased side. In such cases some authors advocate that left sided hepatectomy should be performed and the stones in right lobe should be cleared later by cholangioscopic lithotomy through T-tube tract constructed during surgery or by PTCSL [26,27].

Although intraoperative toilette of remnant liver or transhepatic clearance could also be done, hepaticojunctional anastomosis (HJA) with resection of atrophic hepatic segments seems to be the best option. HJA should also be done in cases with lithiasis affecting both intra and extrahepatic ducts, as this allows the extrahepatic biliary system with its strictures to be bypassed, reducing the risk of recurrent stones. Long term success rates have been achieved by this operation [28].

Satisfactory results have been reported with liver transplantation in selected patients with intrahepatic stones involving both lobes with extensive, severe biliary strictures and bilateral liver atrophy which fail to be treated by other methods [29,30]. Liver transplant seems to be the last choice for OCH with end stage liver disease, however with modern investigations, imaging and systematic management few patients if any should reach this stage of disease.

Extra corporeal shock wave lithotripsy (ESWL) is indicated for cholesterol hepatolithiasis but pre procedural transhepatic cholangioscopic drainage is mandatory for avoiding potential risk of suppurative cholangitis from stone fragment impaction in CBD [31,32]. Its role in RPC is not well established.

#### 6. Our institutional experience

In the Indian subcontinent ascariasis is highly endemic in Kashmir (70%), Bangladesh (82%), central and south west India (20–49%) [33]. A significant number of these patients in Kashmir develop biliary ascariasis. Furthermore up to 5% of these will progress to develop features of OCH within two years [34]. As such a significant proportion of patients admitted to the departments of gastroenterology and surgery in Kashmir have OCH. The majority of these patients need surgery during the progression of the disease and over the last 26 years the department of surgery has gained wide experience in the surgical management of the disease. We have developed and refined our own classification system to grade the patients with OCH who need surgery.

Grade 1 Disease limited to the extrahepatic ducts with stones/worms, No Liver parenchymal disease, no strictures, and CBD size less than 1.5 cm.

Grade 2 Disease limited to the extrahepatic ducts with stones/worms, No liver parenchymal disease, with strictures of CBD, or CBD size more than 1.5 cm.

Grade 3 Disease involving intrahepatic ducts right or left, with stones/worms with dilatable strictures and no liver parenchymal disease.

Grade 4 Disease involving intrahepatic ducts right or left, with stones/worms with severe non-dilatable strictures or parenchymal disease on same side.

Grade 5 Disease involving both intrahepatic ducts, with stones/worms with severe non-dilatable strictures or parenchymal disease on both side.

This classification system has allowed us to assess different surgeries for same grade of disease to determine which one affords better results. Over a period of many years what we found that with lesser or conventional surgery most of the patients come back with recurrent stones and redo surgeries in subsequent sittings are more difficult and more demanding with worse outcomes. Based on our

retrospective experience we now choose the following surgeries for various grades of the disease (See Figs. 1–5 for better understanding of surgeries). Basic modality of management continues to be ERCP; however, the following surgeries are only carried on the patients who are referred to our department after failed ERCP trial or advanced grades of OCH. MRCP has now been an excellent addition in the preoperative workup for better understanding the extent of disease and for better grading and planning for surgery and even for differentiating benign and malignant obstructions. [35]

Grade 1 CBD exploration with T Tube drainage.

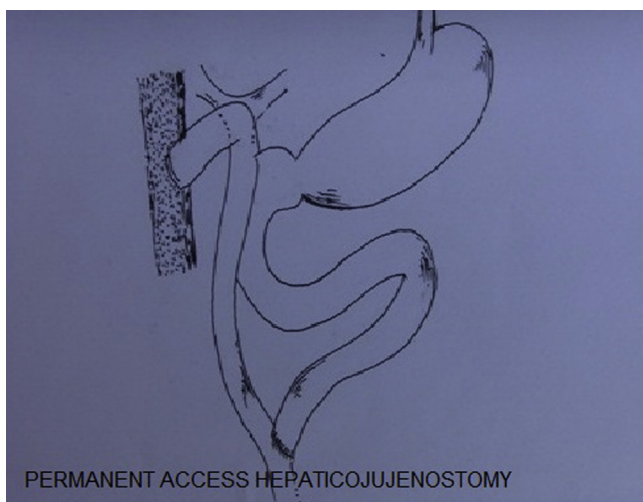
Grade 2 CBD exploration with Hepaticojunostomy with or without access loop

Grade 3 CBD exploration with clearance of intrahepatic stones and dilatation of strictures with Hepaticojunostomy with an access loop.

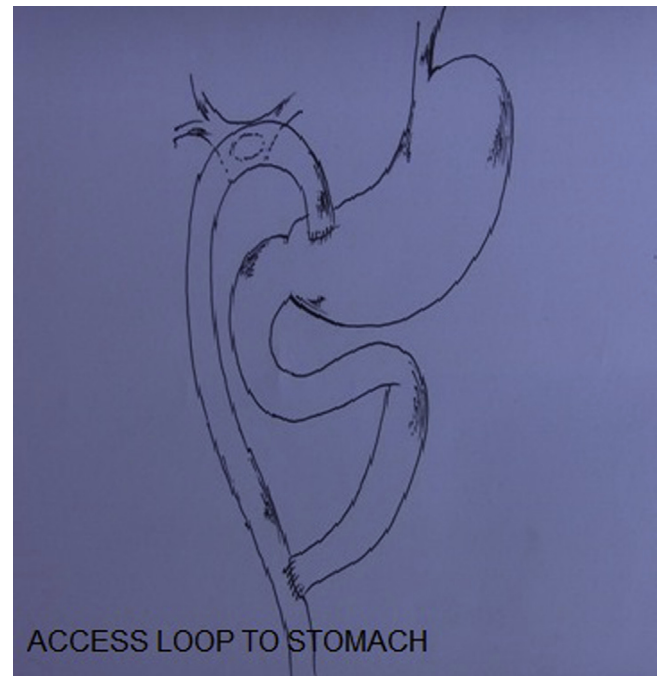
Grade 4 Liver resections, left lateral sectorectomy, Right or Left Hepatectomy.

Grade 5 Hepatectomy on more effected side with clearance of contralateral ducts and dilatation of the strictures and hepaticojunostomy with access loop. Or referral for Liver transplant.

Patients with grade 1 disease have mild ductal changes so they are managed as choledocholithiasis, with confirmation of clearance with choledochoscopy before T Tube drainage. Previously patients with Stage 2 disease were considered candidates for choledochoduodenostomy but our experience has shown that the CBD in these patients is fibrotic and does not drain well so these patients tend to return with recurrent stones above the anastomosis. To avoid recurrent surgeries in these patients we prefer to perform a high biliary enteric anastomosis at the first chance. The choice of access loop in patients with grade 3 disease is decided at the time of surgery depending on local findings. We prefer an internal access loop with the 2nd part of the duodenum in patients being operated for the first time. While in patients who have had previous surgery, we may occasionally opt for an external access limb as it is technically easier and avoids difficult dissection in areas with adhesions. Liver resection in our opinion is the best option for patients with severe strictures and stones impacted proximal to them or for diseased atrophic liver segments. In our setting this commonly

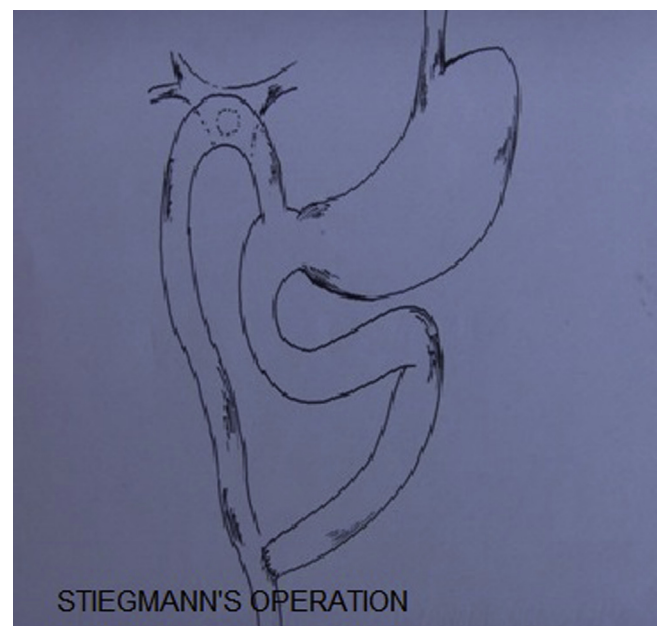


**Fig. 1.** Permanent Access Hepaticojunostomy – a closed internal access loop of jejunum marked with staples put under skin in parietal wall; to be used by an intervention radiologist in case of need to avoid a major surgery.

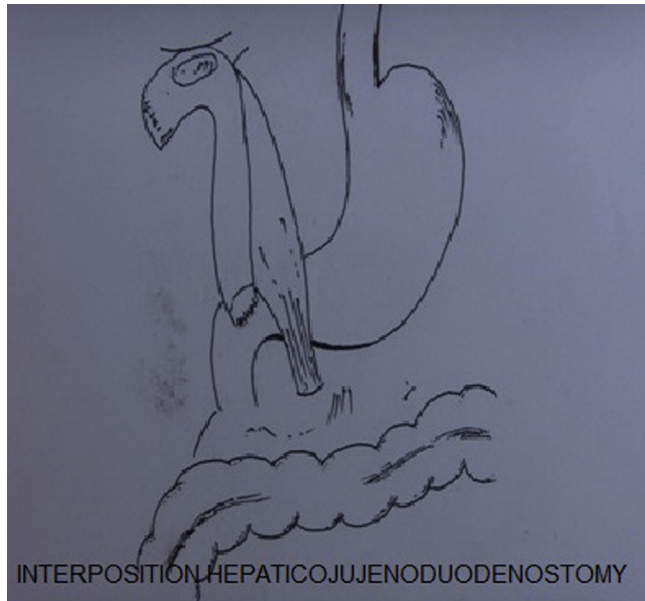


**Fig. 2.** Access Loop to Stomach – an internal access loop of jejunum for future access to an endoscopist to remove the block; not a very comfortable angle for the endoscopist to negotiate.

means a left lateral sectorectomy or a left lobectomy and only a few of our patients require right sided liver resections. Literature also supports hepatectomy for complex and advanced cases to decrease the chances of redo surgeries and recurrent stones [36,37]. Patients with grade 5 disease are most difficult to treat and results are also dissatisfying. Our choice is usually a liver resection on the worse affected side with construction of access loop for future management of recurrent stones on the contralateral side. However in patients with severe bilateral involvement and end stage liver we prefer to refer them for possible liver transplant as the procedure is not performed at our hospital.



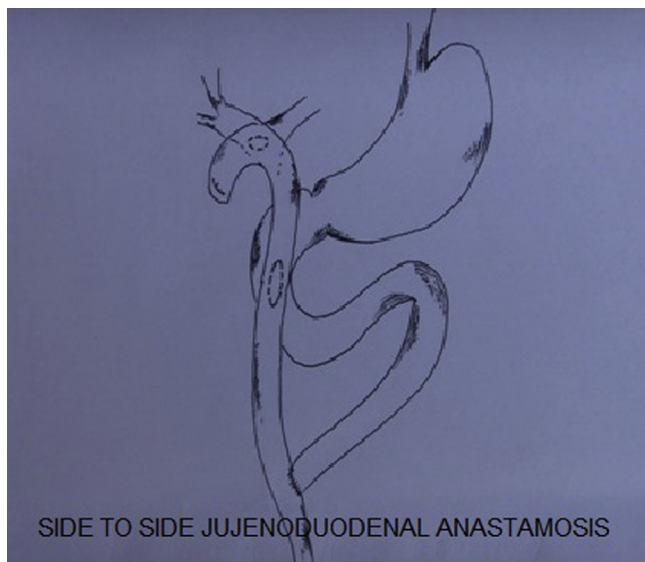
**Fig. 3.** Steigmann's Operation – an internal access loop with proximal duodenum; again not an endoscopist friendly angle.



**Fig. 4.** Interposition Hepaticojujenoduodenostomy – a free pedicled internal access loop of jejunum; time consuming procedure, not with any significant advantage.

The best outcome in patients with OCH can be expected at advanced tertiary care be achieved by following certain principles;

1. Full preoperative evaluation with stress both on extent of biliary tract involvement and liver atrophy.
2. Preoperative counseling of patients about disease with guarded prognosis and need for recurrent interventions.
3. Referral to experienced center if possible.
4. Selection of most appropriate surgical intervention based on accepted guidelines.
5. Frequent postoperative chemotherapy to control parasitic infection.
6. Proper follow up to avoid patients progressing advanced stage of disease.



**Fig. 5.** Side to side Jukenoduodenal anastomosis – most comfortable internal access loop in the second part of duodenum; most convenient endoscopist friendly angle to negotiate; preferred in our set up.

## Ethical approval

Review article, doesn't need any ethical approval.

## Author contribution

Dr Fazl Q Parray conceived the basic idea of manuscript and wrote the basic draft.

Dr Mehmood A Wani, collected all the material required for writing the review and wrote the final draft.

Prof Nazir A Wani gave the final touch to the manuscript.

## Funding

Nil.

## Conflict of interest

No conflict of interest to be declared.

## References

- [1] J.H. Lim, Oriental cholangiohepatitis: pathological, clinical and radiological features, *Am. J. Roentgenol.* 157 (1991) 1–8.
- [2] K. Uchiyama, H. Onishi, M. Tani, H. Kinoshita, M. Ueno, H. Yamaue, Indication and procedure for treatment of hepatolithiasis, *Assrch. Surg.* 137 (2002) 149–153.
- [3] K. Digby, Common duct stones of liver origin, *Br. J. Surg.* 17 (1930) 578–591.
- [4] H.W. Harris, Z.L. Kumwenda, S.M. Sheen-Chen, A. Shah, W.P. Schechter, Recurrent pyogenic cholangitis, *Am. J. Surg.* 176 (1998) 34–37.
- [5] P. Herman, T. Bacchella, V. Pugliese, A.L. Montagnini, M.A. Machado, J.E. da Cunha, et al., Non oriental primary intrahepatic lithiasis: experience with 48 cases, *World J. Surg.* 29 (7) (2005) 858–862.
- [6] S. Kim, S. Moon, B.M. Popkin, The nutrition transition in South Korea, *Am. J. Clin. Nutr.* 71 (2000) 44–53.
- [7] Y.H. Park, S.J. Park, J.Y. Jang, Y.J. Ahn, Y.C. Park, Y.B. Yoon, et al., Changing patterns of gall stone disease in Korea, *World J. Surg.* 28 (2) (2004) 206–210.
- [8] M.S. Khuroo, M.Y. Dar, G.N. Yattoo, Serial cholangiographic appearances in recurrent pyogenic cholangitis, *Gastrointest. Endosc.* 39 (5) (1993) 674–679.
- [9] M. Tabata, F. Nakayama, Bacteriology of hepatolithiasis, *Prog. Clin. Biol. Res.* 152 (1984) 163–174.
- [10] S. Sheen-Chen, W. Chen, H. Eng, C. Sheen, C. Chou, Y. Cheng, T. Lee, Bacteriology and antimicrobial choices in hepatolithiasis, *Am. J. Infect. Control* 28 (2000) 298–301.
- [11] Y. Zen, K. Harada, M. Sasaki, K. Tsuneyama, K. Katayanagi, Y. Yamamoto, et al., Lipopolysaccharide induced over expression of MUC2 and MUC5AC in cultured biliary epithelial cells: possible key phenomenon of hepatolithiasis, *Am. J. Pathol.* 161 (2002) 1475–1484.
- [12] M.J. Zinner, S.I. Schwartz, H. Ellis, Recurrent pyogenic cholangitis, in: J. Wong, S.T. Fan (Eds.), *Maingot's Abdominal Operations*, Appleton and Lange, London, 1997, pp. 1771–1787.
- [13] S. Chou, C.W. Chan, Recurrent pyogenic cholangitis: a necropsy study, *Pathology* 12 (3) (1980) 415–428.
- [14] F. Nakayama, A. Koga, Hepatolithiasis: present status, *World J. Surg.* 8 (1) (1984) 9–14.
- [15] M.H. Kim, J. Sekijima, S.P. Lee, Primary intrahepatic stones, *Am. J. Gastroenterol.* 90 (4) (1995) 540–548.
- [16] T.F. Khan, M.N. Norazmi, Recurrent pyogenic cholangitis: is immune depression a feature? (Letter), *Trop. Doct.* 28 (3) (1998) 187.
- [17] S.T. Fan, T.K. Choi, C.M. Lo, F.P. Mok, E.C. Lai, J. Wong, Treatment of hepatolithiasis: improvement of result by systematic approach, *Surgery* 109 (4) (1991) 474–480.
- [18] C.J. Hung, P.W. Lin, Role of right hepatic lobectomy in the treatment of isolated right sided hepatolithiasis, *Surgery* 121 (1997) 130–134.
- [19] D.H. Bonnel, C.E. Lihuory, F.E. Cornud, J.F. Lefebvre, Common bile duct and intrahepatic duct stones: results of transhepatic electrohydraulic lithotripsy in 50 patients, *Radiology* 180 (1991) 345–348.
- [20] M. Sato, Y. Watanabe, S. Horiuchi, Y. Nakata, N. Sato, Y. Kashu, et al., Long term results of hepatic resections for hepatolithiasis, *HPB Surg.* 9 (1) (1995) 37–44.
- [21] K. Otani, S. Shimizu, K. Chijiwa, T. Ogawa, T. Morisaki, A. Sugitani, et al., Comparison of treatments for hepatolithiasis: hepatic resection versus cholangioscopic lithotomy, *J. Am. Coll. Surg.* 189 (1999) 177–182.
- [22] K.S. Jeng, I. Ohta, F.S. Yang, Reappraisal of the systematic management of complicated hepatolithiasis with bilateral intrahepatic biliary strictures, *Arch. Surg.* 131 (1996) 141–147.
- [23] M.F. Chen, Y.Y. Jan, C.S. Wang, T.L. Hwang, L.B. Jeng, S.C. Chen, et al., Role of hepatic resections in surgery for bilateral intrahepatic stones, *Br. J. Surg.* 84 (9) (1997) 1229–1232.

- [24] Y.H. Yeh, M.H. Huang, J.C. Yang, L.R. Mo, J. Lin, S.K. Yueh, Percutaneous trans hepatic cholangioscopy and lithotripsy in the treatment of intra hepatic stones: a study with 5 year follow up, *Gastrointest. Endosc.* 42 (1) (1995) 13–18.
- [25] K. Chijiwa, N. Kameoka, M. Komura, T. Yamasaki, H. Noshiro, K. Nakano, Hepatic resection for hepatolithiasis and long term results, *J. Am. Coll. Surg.* 180 (1) (1995) 43–48.
- [26] Y.Y. Jan, M.F. Chen, Percutaneous tran hepatic cholangioscopic lithotomy for hepatolithiasis: long term results, *Gastrointest. Endosc.* 42 (1995) 1–5.
- [27] K.S. Jeng, I. Ohta, F.S. Yang, T.P. Liu, S.C. Shih, W.S. Chang, et al., Coexisting sharp ductal angulation with intrahepatic biliary structures in right hepatolithiasis, *Arch. Surg.* 129 (1994) 1097–1102.
- [28] W.H. Nealon, F. Urrutia, Long term follow up after bilioenteric anastomosis for benign bile duct stricture, *Ann. Surg.* 223 (1996) 639–648.
- [29] R.W. Strong, S.P. Chew, D.R. Wall, J. Fawcett, S.V. Lynch, Liver transplantation for hepatolithiasis, *Asian J. Surg.* 25 (2002) 180–183.
- [30] G.D. Pan, L.N. Yan, B. Li, S.E. Lu, Y. Zeng, T.F. Wen, et al., Liver transplantation for patients with hepatolithiasis, *Hepatobiliary Pancreat. Dis. Int.* 4 (2005) 345–349.
- [31] C.D. Becker, J.S. Fache, R.G. Gibney, C.H. Scudamore, H.J. Burhenne, Cholelithiasis: treatment with extra corporeal shock wave lithotripsy, *Radiology* 165 (2) (1987) 407–408.
- [32] K. Uchiyama, H. Tanimura, K. Ishimoto, K. Murakami, T. Nakai, Extra corporeal shock wave lithotripsy for biliary stones: a nation wide in Japan, *Arch. Jpn. Chir.* 63 (1994) 199–207.
- [33] M.S. Khuroo, Ascariasis, *Gastroenterol. Clin. N. Am.* 25 (1996) 553–5570.
- [34] O.J. Shah, S.A. Zargar, I. Robbani, Biliary ascariasis a review, *World J. Surg.* 30 (2006) 1500–1506.
- [35] H.W. Eun, J.H. Kim, S.S. Hong, Y.J. Kim, Malignant versus benign hepatic masses in patients with recurrent pyogenic cholangitis: MR differential diagnosis, *Abdom. Imaging* 37 (5) (Oct 2012) 767–774 (Medline).
- [36] K.F. Lee, C.N. Chong, D. Ng, Y.S. cheung, W. Ng, J. Wong, Outcome of surgical treatment for recurrent pyogenic cholangitis: a single-centre study, *HPB Oxf.* 11 (1) (2009) 75–80 (Medline). (Full Text).
- [37] W.M. Tsui, Y.K. Chan, C.T. Wong, Y.F. Lo, Y.W. Yeung, Y.W. Lee, Hepatolithiasis and the syndrome of recurrent pyogenic cholangitis: clinical, radiologic, and pathologic features, *Semin. Liver Dis.* 31 (1) (Feb 2011) 33–48 (Medline).