**Supplement 1. Checklist of items in reporting this systematic review (PRISMA statement)**

|  |  |  |  |
| --- | --- | --- | --- |
| **Section/topic** | **Item**  | **Checklist item** | **Reported** **on page**  |
| **Title** |
| Title | 1 | Identify the report as a systematic review, meta-analysis, or both | 1 |
| **Abstract** |  |  |  |
| Structured summary | 2 | Provide a structured summary including, as applicable, background, objectives, data sources, study eligibility criteria, participants, interventions, study appraisal and synthesis methods, results, limitations, conclusions and implications of key findings, systematic review registration number | 2 |
| **Introduction** |
| Rationale | 3 | Describe the rationale for the review in the context of what is already known | 4 |
| Objectives | 4 | Provide an explicit statement of questions being addressed with reference to participants, interventions, comparisons, outcomes, and study design (PICOS) | 5 |
| **Methods** |
| Protocol and registration | 5 | Indicate if a review protocol exists, if and where it can be accessed (such as web address), and, if available, provide registration information including registration number | N/A |
| Eligibility criteria | 6 | Specify study characteristics (such as PICOS, length of follow-up) and report characteristics (such as years considered, language, publication status) used as criteria for eligibility, giving rationale | 5-6 |
| Information sources | 7 | Describe all information sources (such as databases with dates of coverage, contact with study authors to identify additional studies) in the search and date last searched | 5 |
| Search | 8 | Present full electronic search strategy for at least one database, including any limits used, such that it could be repeated | 5 |
| Study selection | 9 | State the process for selecting studies (that is, screening, eligibility, included in systematic review, and, if applicable, included in the meta-analysis | 5-6 |
| Data collection process | 10 | Describe method of data extraction from reports (such as piloted forms, independently, in duplicate) and any processes for obtaining and confirming data from investigators | 6 |
| Data items | 11 | List and define all variables for which data were sought (such as PICOS, funding sources) and any assumptions and simplifications made | 6 |
| Risk of bias in individual studies | 12 | Describe methods used for assessing risk of bias of individual studies (including specification of whether this was done at the study or outcome level), and how this information is to be used in any data synthesis | 7 |
| Summary measures | 13 | State the principal summary measures (such as risk ratio, difference in means) | 7 |
| Synthesis of results | 14 | Describe the methods of handling data and combining results of studies, if done, including measures of consistency (such as I2 statistic) for each meta-analysis | 7 |
| Risk of bias across studies | 15 | Specify any assessment of risk of bias that may affect the cumulative evidence (such as publication bias, selective reporting within studies) | 7 |
| Additional analyses | 16 | Describe methods of additional analyses (such as sensitivity or subgroup analyses, meta-regression), if done, indicating which were pre-specified | N/A |
| **Results** |
| Study selection | 17 | Give numbers of studies screened, assessed for eligibility, and included in the review, with reasons for exclusions at each stage, ideally with a flow diagram | 8 |
| Study characteristics | 18 | For each study, present characteristics for which data were extracted (such as study size, PICOS, follow-up period) and provide the citations | 9 |
| Risk of bias within studies | 19 | Present data on risk of bias of each study and, if available, any outcome-level assessment (see item 12). | N/A |
| Results of individual studies | 20 | For all outcomes considered (benefits or harms), present for each study (a) simple summary data for each intervention group and (b) effect estimates and confidence intervals, ideally with a forest plot | N/A |
| Synthesis of results | 21 | Present results of each meta-analysis done, including confidence intervals and measures of consistency | N/A |
| Risk of bias across studies | 22 | Present results of any assessment of risk of bias across studies (see item 15) | 15 |
| Additional analysis | 23 | Give results of additional analyses, if done (such as sensitivity or subgroup analyses, meta-regression) (see item 16) | N/A |
| **Discussion** |
| Summary of evidence | 24 | Summarize the main findings including the strength of evidence for each main outcome; consider their relevance to key groups (such as health care providers, users, and policy makers | 16 |
| Limitations | 25 | Discuss limitations at study and outcome level (such as risk of bias), and at review level (such as incomplete retrieval of identified research, reporting bias) | 20 |
| Conclusions | 26 | Provide a general interpretation of the results in the context of other evidence, and implications for future research | 20-21 |
| **Funding** |
| Funding | 27 | Describe sources of funding for the systematic review and other support (such as supply of data) and role of funders for the systematic review | 21 |

N/A: not applicable

Modified from: David Moher, Alessandro Liberati, Jennifer Tetzlaff, Douglas G Altman. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. BMJ 2009;339:b2535.

**Supplement 2. Actual search study of this systematic review**

Ovid

Database(s): Embase 1988 to 2017 Week 31, EBM Reviews - Cochrane Central Register of Controlled Trials June 2017, EBM Reviews - Cochrane Database of Systematic Reviews 2005 to July 26, 2017, Ovid MEDLINE(R) Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present
Search Strategy:

|  |  |  |
| --- | --- | --- |
| **#** | **Searches** | **Results** |
| 1 | exp bile duct cyst/ | 2475 |
| 2 | exp Bile Duct Diseases/ and exp Cysts/ | 8062 |
| 3 | (peribiliary or "peri-biliary" or periductal or "peri-ductal").ti,ab,hw,kw. | 3542 |
| 4 | (1 or 2) and 3 | 126 |
| 5 | ((peribiliary or "peri-biliary" or periductal or "peri-ductal") adj5 (cyst\* or polycyst\* or multicyst\*)).ti,ab,hw,kw. | 168 |
| 6 | 4 or 5 | 207 |
| 7 | remove duplicates from 6 | 126 |

Scopus

1. TITLE-ABS-KEY ( ( ( peribiliary  OR  "peri-biliary"  OR  periductal  OR  "peri-ductal" )  W/5  ( cyst\*  OR  polycyst\*  OR  multicyst\* ) ) )
2. PMID(0\*) OR PMID(1\*) OR PMID(2\*) OR PMID(3\*) OR PMID(4\*) OR PMID(5\*) OR PMID(6\*) OR PMID(7\*) OR PMID(8\*) OR PMID(9\*)
3. 1 and not 2

**Supplement 3**

**METHODS:**

 **Data Sources and Search Strategies:** A comprehensive search of several databases from each database’s inception to August 2nd, 2017, any language was conducted. The databases included Ovid MEDLINE Epub Ahead of Print, Ovid Medline In-Process & Other Non-Indexed Citations, Ovid MEDLINE, Ovid EMBASE, Ovid Cochrane Central Register of Controlled Trials, Ovid Cochrane Database of Systematic Reviews, and Scopus. The search strategy was designed and conducted by an experienced librarian with input from the study’s principle investigator. Controlled vocabulary supplemented with keywords was used to search for peribiliary cysts. The actual strategy is provided in supplement II. In addition, the first 300 entries of Google Scholar were searched using the terms “peribiliary cysts” to look for studies not included in major databases. Reference lists were also manually reviewed for additional cases. The detailed search strategy is in Supplement 2.

 **Inclusions criteria based on the following definitions:**

*Frequency of peribiliary cysts:* This is based on imaging, surgical or postmortem studies that evaluated peribiliary cysts.

 *Diagnosis of peribiliary cysts:* This is done by one or more of the following imaging modalities: abdominal contrast-enhanced computed tomography (CECT), drip infusion cholangiographic computed tomography (DIC-CT), magnetic resonance imaging (MRI), magnetic resonance cholangio-pancreatography (MRCP); or examination of surgical or autopsy specimen.

 *Clinical manifestations of peribiliary cysts:* This is based on case reports and case-series that reported sufficient clinical data

 **Data Extraction and Assessment**

 The data included year of publication; country of origin; publication language; publication format (full-text article, letter to the editors, image in medicine, abstract form); type of study (case report, case-series); age; gender; alcohol consumption; associated conditions or diseases [cirrhosis and etiology, portal hypertension, portal thrombosis, neoplasm, cholangitis, autosomal dominant polycystic kidney disease (ADPKD), autosomal dominant polycystic liver disease (ADPLD)]; clinical manifestations; liver function tests; tumor markers [carcino-embryologic antigen (CEA), carbohydrate antigen 19-9 (CA19-9)]; imaging modalities [abdominal contrast-enhanced computed tomography (CECT), drip infusion cholangiographic computed tomography (DIC-CT), abdominal magnetic resonance imaging (MRI), magnetic resonance cholangio-pancreatography (MRCP), endoscopic ultrasound (EUS), intraductal endoscopic ultrasound (ID-EUS), endoscopic retrograde cholangio-pancreatography (ERCP), percutaneous trans-hepatic cholangiography (PTC), endoscopic or intraoperative cholangioscopy]; accurate or inaccurate diagnosis of the disease; modalities allowing to make the diagnosis (imaging modalities, surgery or autopsy); number, size, and location of the cysts along the biliary tract; complications related to the disease; duration of follow-up after diagnosis and whether there was an increase in number and/or size of the cysts; and lastly final outcome, cause of death and postmortem exam.

 **Assessment of methodological qualities of case reports and case series:**

 Given that there are no available validated tools to assess the methodological quality of case reports and case series, we derived items from the Newcastle-Ottawa Scale (NOS) that were appropriate for this systematic review. We removed from the NOS the items that relate to comparability and adjustment (because the studies included were non-comparative). We retained for the purpose of methodological quality the items that focused on selection, representativeness of cases, and ascertainment of outcome and exposure This resulted in 5 criteria in the form of questions with a binary response (yes/no), whether the item was suggestive of bias or not. These questions are listed in Supplement 4. We considered the quality of the report good when all 5 criteria were fulfilled, moderate when 4 were fulfilled, and low when ≤3 were fulfilled.

 **RESULTS:**

 The flow diagram through the different phases of this systematic review is shown in Supplement 5. We identified 9 studies that assessed the frequency and 72 case reports and case series with sufficient clinical data [two of them were included to assess the frequency ([1](#_ENREF_1), [2](#_ENREF_2))].

 **Studies characteristics:**

Seventy-twocase reports and case series from 10 countries published between 1984 and 2016 with sufficient clinical data were included. We identified 48 studies through the librarian search, 17 studies through manual search of reference lists ([3-19](#_ENREF_3)) and 7 studies through the Google Scholar database ([20-26](#_ENREF_20)). The publication language was Korean in 3 studies ([10](#_ENREF_10), [16](#_ENREF_16), [27](#_ENREF_27)), Japanese in 20 studies ([4-6](#_ENREF_4), [8](#_ENREF_8), [9](#_ENREF_9), [11-14](#_ENREF_11), [17](#_ENREF_17), [18](#_ENREF_18), [21-24](#_ENREF_21), [26](#_ENREF_26), [28-31](#_ENREF_28)), and English in the remaining studies. One publication was in an abstract form ([32](#_ENREF_32)), two were in form of letters to the editor ([33](#_ENREF_33), [34](#_ENREF_34)), four in form of image in medicine ([35-38](#_ENREF_35)) and all others were full-text manuscripts. There were 56 case reports and 16 case series that included 2 to 13 patients ([1-3](#_ENREF_1), [29-31](#_ENREF_29), [39-48](#_ENREF_39)). Supplement 7 demonstrates the number of reported cases from different countries. Most studies were reported from Japan [Japanese studies: 54/72 studies (75%) – Japanese cases 88/135 (65%)].

 We excluded one duplicate study (Kim 2013) and 10 studies with insufficient clinical data some of them were duplicated (Qian 2003, Itai 2003, Brancatelli 2007, Matsubara 2012, Galia 2014, Matsubara 2014, Sato 2014, Katabathina 2016, Ronot 2017, Kozaka 2017). We excluded three patients with non-communicating cysts observed after Kasai’s operation for biliary atresia that were thought to be related to peribiliary glands without confirmation (Kawarasaki 1997), eight patients in the study of Colina et al. diagnosed as mucoceles of the cystic duct remnant in liver transplant patients (Colina 1999), and one patient with cyst in the wall of common bile duct that was thought to be related to a hematoma from the stump of the cystic artery during cholecystectomy (Scotiniotis 2001). References of excluded studies are found in supplement 8.

**Supplement 4**

|  |
| --- |
| **Tool for methodological qualities assessment of case reports and case series** |
| 1. Did the patient(s) represent the whole case(s) of the medical center?  |
| 2. Was the diagnosis correctly made?  |
| 3. Were other important diagnosis excluded?  |
| 4. Were all important data cited in the report?  |
| 5. Was the outcome correctly ascertained?  |

**Supplement 5**

The flow diagram through the different phases of this systematic review



**Supplement 6. References of excluded studies**

|  |  |  |
| --- | --- | --- |
| **References** | **No of patients**  | **Reason for exclusion** |
| **Duplicate study** |
| Kim H. Peribiliary cyst in healthy young patient. HPB 2013, 15 (Suppl. 2), 90A. | 1 patient | – |
| **Insufficient clinical data** |
| Qian Q, Li A, King BF, Kamath PS, Lager DJ, Huston J, et al. Clinical profile of autosomal dominant polycystic liver disease. Hepatology 2003;37(1):164–71. | – | study of autosomal dominant polycystic liver disease |
| Itai Y. Peribiliary cysts. Shoukaki Gazou (Journal of Gastroenterological Imaging). 2003; 5:35-40. | – | imaging study of peribiliary cysts |
| Brancatelli G, Federle MP, Ambrosini R et al. Cirrhosis: CT and MR imaging evaluation. European Journal of Radiology 2007;61: 57–69. | 1 patient | revue article of imaging for peribiliary cysts |
| Takashi Matsubara, Yasunori Sato, Motoko Sasaki, et al. Immunohistochemical characteristics and malignant progression of hepatic cystic neoplasms in comparison with pancreatic counterparts. Human Pathology 2012; 43, 2177–2186. | 10 patients | histological study without clinical details |
| Massimo Galia, Adele Taibbi, Daniele Marin et al. Focal lesions in cirrhotic liver: what else beyond hepatocellular carcinoma? Diagn Interv Radiol 2014; 20:222-228. | 1 patient | review article of imaging for peribiliary cysts |
| Matsubara T, Sato Y, Igarashi S et al. Alcohol-related injury to peribiliary glands is a cause of peribiliary cysts: based on analysis of clinical and autopsy cases. J Clin Gastroenterol 2014 Feb;48(2):153-9. | 60 patients |  |
| Yasunori Sato, Kenichi Harada, Motoko Sasaki, Yasuni Nakanuma? Cystic and micropapillary epithelial changes of peribiliary glands might represent a precursor lesion of biliary epithelial neoplasms. Virchows Arch 2014;464:157-163. | 49 patients | Histologic study of cystic change of peribiliary cysts (> 2 mm) |
| Katabathina VS, Flaherty EM, Dasyam AK et al. Biliary Diseases with Pancreatic Counterparts”: Cross-sectional Imaging Findings. RadioGraphics 2016; 36:374–392. | 3 patients | imaging study |
| Maxime Ronot, Marco Dioguardi Burgio, Yvonne Purcell, Romain Pommier, Giuseppe Brancatelli, Valerie Vilgrain. Focal lesions in cirrhosis: not always HCC. European Journal of Radiology 2017:93:157-168. | 1 patient  | review article of focal lesions in cirrhosis  |
| Kazuto Kozaka and Osamu Matsu. Pathology and Imaging of Peribiliary Cysts: Recent Progress. In: Y. Nakanuma (ed.), Pathology of the Bile Duct, Springer, Singapore, 2017, pp 113-120. | 2 patients | revue article of imaging for peribiliary cysts |
| Kawarasaki H, Itoh M, Mizuta K, Tanaka H, Makuuchi M. Further observations on cystic dilatation of the intrahepatic biliary system in biliary atresia after hepatic portoenterostomy: report on 10 cases. Tohoku J. Exp. Med.1997; 181: 175–83. | 3 patients | cysts after Kasai operation for biliary atresia |
| Colina F, Castellano VM, Gonzalez-Pinto I, et al. Hilar biliary cysts in hepatic transplantation. Report of three symptomatic cases and occurrence in resected liver grafts. Transpl Int 1998; 11:110–116. | 8 patients | mucoceles of cystic duct remnant in liver transplant patients |
| **Diagnosis of peribiliary cysts not confirmed** |
| Scotiniotis IA, Kochman ML. Intramural cyst of the bile duct demonstrated by cholangioscopy and intraductal US. Gastrointestinal Endoscopy 2001;54 (2): 260-262. | 1 patient | thought to be related to bleeding during cholecystectomy |
| **Total: 14 studies** |  |  |

**Supplement 7. Countries of reported studies of peribiliary cysts**

|  |  |  |  |
| --- | --- | --- | --- |
| **Countries** | **No of studies** | **No of reported cases** | **References** |
| Japan | 54 | 88 | Nakanuma 1984, Kasai 1984, Ueki 1987, Yoshinaga 1989, Terada 1992, Itai 1994, Terayama 1995, Akiyama 1997, Yuasa 1997, Terasaki 1997, Ishihara 1997, Fujioka 1997, Scolari 1997, Ahmadi 1997, Nagata 1998, Nishimura 1998, Takakura 2000, Miyake 2001, Motoo 2001, Okada 2001, Chiba 2002, Murai 2002, Cho 2003, Ohta 2003, Terada 2003, Kolodziejski 2004, Seguchi 2004, Shigeta 2005, Tsuchiya 2005, Yasutomo 2005, Yokomichi 2006, Otani 2006, Miura 2006, Ohnishi 2007, Kai 2008, Hamasu 2008, Ikenaga 2009, Tohma 2009, Nakanishi 2009, Fujiwara 2009, Sato 2009, Takahashi 2009, Nakayama 2010, Matsukura 2010, Terada 2010, Nakanishi 2011, Takuma 2012, Zen 212, Kozuki 2012, Teramoto 2015, Shibata 2015, Aoba 2015, Umemura 2016, Iijima 2016 |
| South Korea | 5 | 5 | Lee 1999, Park 2001, Kim 2009, Seo 2012, Kim 2013 |
| India | 2 | 14 | Gupta 1999, Johnson 2007 |
| Taiwan | 1 | 1 | Su 2013 |
| Australia | 1 | 2 | Pang 2010 |
| Spain  | 1 | 5 | Colina 1998 |
| France | 2 | 2 | Da Ines 2011, Montoriol 2012 |
| UK | 1 | 1 | Fusai 2005 |
| USA | 4 | 15 | Herman 1990, Baron 1994, Stevens 1996, Lim 2016 |
| Canada | 1 | 2 | Wanless 1987 |
| **Total: 10 countries** | **72** | **135** |  |

**Supplement 8. Frequency of peribiliary cysts in imaging, surgical and postmortem studies**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Author/Year** | **Country** | **Underlying condition** | **Diagnostic modality** | **No of patients** | **No of PBCs** (%) | **Size of cysts**mm |
| Terada/1990 ([49](#_ENREF_49)) | Japan | Postmortem | autopsy | 1000 | 202 (20.2%) | most microscopic bile ducts compression in 16  |
| Kida/1992 ([50](#_ENREF_50)) | Japan | ADPLD | autopsy | 8 | 8 (100%) | all macroscopic |
| solitary nonparasitic cyst | 18 | 13 (72%) | 11microscopic-2macroscopic |
| normal liver | 23 | 8 (35%) | all microscopic |
| Itai/1995 ([51](#_ENREF_51)) | Japan | ADPKD & their relatives | CT or CECT | 64 | 22 (34%) | ˂10 mm - 30 mm in 1 patient |
| Hoshiba/1996 ([52](#_ENREF_52)) | Japan | liver cirrhosis | CT or CECT | 346 | 31 (9%) | ˂ 15 mm |
| no liver cirrhosis | 307 | 10 (3%) | ˂ 15 mm |
| Colina/1998 ([1](#_ENREF_1)) | Spain | liver transplant patients | resected graft/ autopsy | 493 | 5 (1%) | 5-20 mm |
| Gupta/1999 ([2](#_ENREF_2)) | India | ADPKD | CECT | 24 | 13 (54%) | 10 mm (some 10-30 mm)  |
| Dranssart/2002 ([53](#_ENREF_53)) | France | ADPKD | MRCP  | 90 | 59 (66%) | not reported |
| Matsubara 2014 ([54](#_ENREF_54)) | Japan | heavy alcohol use | autopsy | 202 | 29 (14%) | several mm to 20 mm |
| Goossens/2017 ([55](#_ENREF_55)) | Switzerland | liver transplant patients | resected graft | 71 | 17 (24%) | > 1 mm |
| **Total: 9 studies** | **5 countries** |  |  | **2646** | **417 (16%)** | **1-30 mm** |

ADPKD: autosomal dominant polycystic kidney disease – ADPLD: autosomal dominant polycystic liver disease – CECT: contrast enhanced computed tomography – CT: computed tomography – MRCP: magnetic resonance cholangiopancreatography – PBCs: peibiliary cysts.

**Supplement 9. Type of neoplasms associated with peribiliary cysts based on 135 cases**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Type of neoplasms** | **No of neoplasm**  | **Past history of neoplasm** | **Actual neoplasm** | **References** |
| Hepatocellular carcimoma | 16 | –  | 16 | Nakanuma 1984, Wanless 1987, Terada 1992, Baron 1994, Terayama 1995, Ahmadi 1997, Seguchi 2004, Kai 2008, Tohma 2009, Zen 2012 |
| Intrahepatic cholangiocarcinoma1 | 1 | – | 1 | Otani 2006 |
| Extrahepatic cholangiocarcinoma | 1 | – | 1 | Miura 2006 |
| Intraductal papillary neoplasm of bile duct | 4 | – | 4 | Nakanishi 2009, Nakanishi 2011, Zen 2012 |
| Cancer of head of the pancreas | 1 | – | 1 | Fujiwara 2009 |
| Intraductal papillary neoplasm of pancreas | 1 | 1 | – | Sato 2009 |
| Lung cancer | 1 | – | 1 | Terada 2010 |
| Esophageal cancer | 1 |  | 1 | Takuma 2012 |
| Gastric cancer | 7 | 3 | 4 | Itai 1994, Akiyama 1997, Yuasa 1997, Ohta 2003, Seguchi 2004, Yasutomo 2005, Tohma 2009  |
| Colon cancer | 4 | 3 | 1 | Itai 1994, Nagata 1998, Ohta 2003, Aoba 2015 |
| Prostate cancer | 1 | 1 | – | Kolodziejski 2004 |
| Acute myeloid leukemia | 1 | – | 1 | Takahashi 2009 |
| **Total: 10 types of neoplasms** | **39 neoplasms****37 patients2** | **8** | **31** |  |

(1) diagnosis based on imaging and not confirmed histologically – (2) Two patients had 2 simultaneous neoplasms: hepatocellular carcinoma and early gastric cancer (Seguchi 2004), hepatocellular carcinoma and intraductal papillary neoplasm of the bile duct (Zen 2012).

**Supplement 10.** **Diagnostic modalities of peribiliary cysts based on 135 patients**

|  |  |  |
| --- | --- | --- |
|  | **No of patients**  | **Correct diagnosis**  |
| **Diagnosis without previous imaging modalities**Diagnosis at surgeryDiagnosis at autopsy | **20**911 | – – –  |
| **Performance of one or more imaging modalities**Correct diagnosisIncorrect diagnosis - Diagnosis at surgery - Diagnosis at autopsy | **115**6946379 | – 69/115 (60%)46/115 (40%)––  |
| **Diagnostic accuracy of different imaging modalities**CECTDIC-CTMRIMRCPCholangioscopy1IDUS | 105192039124 | 51**/**105(48%)15/19 (79%)9/20 (45%)25/39 (64%)7/12 (58%)3/4 (75%) |

(1) Cholangioscopy was performed during ERCP or intra-operatively and revealed submucosal lesions – CECT: contrast-enhanced computed tomography – DIC-CT: drip infusion cholangiographic computed tomography – EUS: endoscopic ultrasound – IDUS: intraductal ultrasound – MRCP: magnetic resonance cholangio-pancratography – MRI: magnetic resonance imaging.

**Supplement 11. Methodological quality assessment of 135 cases of peribiliary cysts**

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| **First author/Year** | **No** | **Question 1** | **Question 2** | **Question 3** | **Question 4** | **Question 5** | **Assessment** |
| **yes** | **no** | **yes** | **no** | **yes** | **no** | **yes** | **no** | **yes** | **no** |
| Nakanuma/1984([3](#_ENREF_3)) | 8 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 8 |
| Kasai/1984([4](#_ENREF_4)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Wanless/1987([39](#_ENREF_39)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| Ueki/1987([5](#_ENREF_5)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Yoshinaga/1989([6](#_ENREF_6))  | 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| Herman/1990([56](#_ENREF_56)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Terada/1992([57](#_ENREF_57)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Baron/1994([40](#_ENREF_40)) | 12 | yes |  | yes |  | yes |  | yes |  |  yes |  | good 12 |
| Itai/1994([42](#_ENREF_42)) | 4 | yes |  | yes |  | yes |  | yes |  | yes |  | good 4 |
| Terayama/1995([45](#_ENREF_45)) | 7 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 7 |
| Stevens/1996([58](#_ENREF_58)) | 1 | yes |  | yes |  | yes |  | yes | no | yes |  | moderate 1 |
| Akiyama/1997([7](#_ENREF_7)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Yuasa/1997([59](#_ENREF_59)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Terasaki/1997([44](#_ENREF_44)) | 2 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 2 |
| Ishihara/1997 ([28](#_ENREF_28)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Fujioka/1997([41](#_ENREF_41)) | 3 | yes |  | yes |  | yes |  | yes |  | yes |  | good 3 |
| Scolari/1997([19](#_ENREF_19)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Ahmadi/1997([60](#_ENREF_60)) | 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| Nagata/1998([29](#_ENREF_29)) | 3 | yes |  | yes |  | yes |  | yes |  | yes |  | good 3 |
| Nishimura/1998([8](#_ENREF_8)) | 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| Colina/1998([1](#_ENREF_1)) | 5 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 5 |
| Lee/1999([61](#_ENREF_61)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Gupta/1999([2](#_ENREF_2)) | 13 | yes |  | yes |  | yes |  |  | no |  | no | low 13 |
| Takakura/2000 ([9](#_ENREF_9)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Miyake/2001([35](#_ENREF_35)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Motoo/2001([62](#_ENREF_62)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Okada/2001([63](#_ENREF_63)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Park 2001 ([10](#_ENREF_10)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Chiba/2002([36](#_ENREF_36)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Murai/2002 ([11](#_ENREF_11)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Cho/2003 ([12](#_ENREF_12)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Ohta/2003 ([30](#_ENREF_30)) | 6 | yes |  | yes |  | yes |  | yes |  | yes |  | good 6 |
| Terada/2003([64](#_ENREF_64)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Kolodziejski/2004([20](#_ENREF_20)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Seguchi/2004([43](#_ENREF_43)) | 3 | yes |  | yes |  | yes |  | yes |  | yes |  | good 3 |
| Shigeta/2005 ([21](#_ENREF_21)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Tsuchiya/2005 ([13](#_ENREF_13)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Fusai/2005([65](#_ENREF_65)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Yasutomo/2005 ([14](#_ENREF_14)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Yokomichi/2006([66](#_ENREF_66)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Otani/2006 ([31](#_ENREF_31)) | 4 | yes |  | yes |  | yes |  | yes |  | yes |  | good 4 |
| Miura/2006([67](#_ENREF_67)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Johnson/2007([68](#_ENREF_68)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Ohnishi/2007([22](#_ENREF_22)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Kai/2008([69](#_ENREF_69)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Hamasu/2008 ([23](#_ENREF_23)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Ikenaga/2009([70](#_ENREF_70)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Nakanishi/2009([71](#_ENREF_71)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Tohma/2009([48](#_ENREF_48)) | 3 | yes |  | yes |  | yes |  | yes |  | yes |  | good 3 |
| Fujiwara/2009([34](#_ENREF_34)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Kim/2009([16](#_ENREF_16)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Sato/2009([72](#_ENREF_72)) | 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| Takahashi/2009([33](#_ENREF_33)) | 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| Pang/2010([46](#_ENREF_46)) | 2 | yes |  | yes |  | yes |  | yes |  | yes |  | good 2 |
| Nakayama/2010([37](#_ENREF_37)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Matsukura/2010([24](#_ENREF_24)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Terada/2010([73](#_ENREF_73)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Da Ines/2011([38](#_ENREF_38)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Nakanishi/2011([15](#_ENREF_15)) | 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| Takuma/2012 ([18](#_ENREF_18)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Montoriol/2012([74](#_ENREF_74)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Zen/2012([47](#_ENREF_47)) | 1 | yes |  | yes |  | yes |  |  | no | yes |  | moderate 1 |
| 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Kozuki/2012 ([17](#_ENREF_17)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Seo/2012 ([27](#_ENREF_27)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Su/2013([25](#_ENREF_25)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Kim /2013([75](#_ENREF_75)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Teramoto /2015([32](#_ENREF_32)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Shibata/2015([76](#_ENREF_76)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Aoba/2015 ([26](#_ENREF_26)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Umemura/2016([77](#_ENREF_77)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Iijima/2016([78](#_ENREF_78)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| Lim/2016([79](#_ENREF_79)) | 1 | yes |  | yes |  | yes |  | yes |  | yes |  | good 1 |
| **Total: 72 studies** | 135 | 135 | 0 | 135 | 0 | 135 | 0 | 91 | 44 | 122 | 13 | good 91 (67%)moderate 31 (23%)low 13 (10%) |

**Supplement 12. Characteristic of peribiliary cysts according to their location along the biliary tract**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Characteristics** | **Type I** Intrahepatic118 patientsAge 63 (4-88)1 | **Type II** Extrahepatic9 patientsAge 61 (43-79)1 | **Type III** Intra- & extrahepatic8 patientsAge 70.5 (53 -77)1 | **p value**  |
| Males (%) | 89/105 (85%) | 2/9 (22%) | 6/8 (75%) | 0.001 |
| Japanese patients (%) | 74/118 (63%) | 5/9 (55%) | 6/8 (75%) | 0.784 |
| Associated conditions or diseases (%) | 104/118 (88%) | 6/9 (66%) | 7/8 (87.5%) | 0.168 |
| Cirrhosis (%) | 49/118 (41%) | 0/9 (0%) | 3/8 (37.5%) | 0.033 |
| Portal hypertension (%) | 37/101 (37%) | 0/9 (0%) | 3/7 (43%) | 0.046 |
| Hepatic, biliary or pancreatic neoplasms (%) | 23/118 (19.5%) | 0//9 (0%) | 0/8 (0%) | 0.197 |
| Incorrect diagnosis by imaging modalities (%) | 38/104 (36%) | 5/6 (83%) | 4/5 (80%) | 0.012 |
| Solitary cysts (%) | 4/118 (3.4%) | 9/9 (100%) | 0/8 (0%) | 0.001 |
| Dilated biliary tract (%) | 38/111 (34%) | 4/9 (44%) | 5/6 (83%) | 0.057 |
| Obstructive jaundice (%) | 12/111 (11%) | 4/9 (44%) | 2/6 (33%) | 0.012 |
| Cholangitis (%) | 12/89 (13%) | 1/9 (11%) | 1/6 (17%) | 1.000 |
| Surgical interventions (%) | 49/118 (41.5%) | 8/9 (89%) | 4/8 (50%) | 0.019 |
| Therapeutic misadventures (%) | 18/49 (37%) | 1/8 (12.5) | 1/4 (25%) | 0.517 |
| Mortality rate (%) | 25/105 (24%) | 1/9 (11%) | 3/8 (37.5%) | 0.461 |

(1) Median age and range.

**Supplement 13. Comparison between multiple versus solitary cysts**

|  |  |  |  |
| --- | --- | --- | --- |
| **Characteristics** | **Multiple cysts** 122 patientsAge 64 (4-88)1 | **Solitary cysts** 13 patientsAge 61 (9-81)1 | **p value** |
| Males (%) | 94/109 (86%) | 4/13 (31%) | 0.0027 |
| Japanese patients (%) | 78/122 (64%) | 7/13 (54%) | 0.474 |
| Associated conditions or diseases (%) | 109/122 (89%) | 6/13 (46%) | 0.0001 |
| Cirrhosis (%) | 51/122 (42%) | 1/13 (8%) | 0.0166 |
| Portal hypertension (%) | 40/106 (38%) | 0/11 (0%) | 0.0152 |
| Hepatic, biliary or pancreatic neoplasms (%) | 23/122 (19%) | 0/13 (0%) | 0.2180 |
| Incorrect diagnosis by imaging modalities (%) | 40/107 (37%) | 7/8 (87%) | 0.0078 |
| Dilated biliary tract (%) | 42/113 (37%) | 5/13 (38%) | 0.9272 |
| Obstructive jaundice (%) | 14/113 (12%) | 4/13 (31%) | 0.2306 |
| Cholangitis (%) | 12/92 (13%) | 1/12 (8%) | 1.0000 |
| Surgical interventions (%) | 49/122 (40%) | 12/13 (92%) | 0.2097 |
| Therapeutic misadventures (%) | 19/49 (39%) | 1/ 12 (8%) | 0.0826 |
| Mortality rate (%) | 28/109 (26%) | 1/13 (8%) | 0.1878 |

(1) Median age and range.

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