Journal of Pediatric Surgery Open 4 (2023) 100092

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Open

journal homepage: www.elsevier.com/locate/yjpso

# Association between duodenal atresia/stenosis and biliary and pancreatic abnormalities: Is it overlooked? A systematic review

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ARTICLE INFO

*Keywords:*
Biliary anomalies
Pancreatic anomalies
Biliary atresia
Choledochal cyst
Annular pancreas
Anomalous bifurcated bile duct
Duodenal atresia
Duodenal stenosis
Duodenal obstruction

ABSTRACT

*Background:* Congenital duodenal obstruction occur with other anomalies in 50% of cases. Although predictable, its association with biliary/pancreatic anomalies is rarely reported. All cases of duodenal atresia (DA) and stenosis (DS) associated with biliary and/or pancreatic abnormalities were analyzed.
*Methods:* A systematic review searched all studies reporting association between DA/DS and biliary/pancreatic abnormalities. The study was performed according to PRISMA guidelines.
*Results:* Of 522 studies reviewed, 285 were analyzed after removing the duplicates, and 104 (249 patients) meeting the inclusion criteria, were included. Anomalous bifurcated bile duct was described in 24 cases (20 DA, 4 DS): 6 at necropsy, and 20 at perinatal surgery. Choledochal cyst was found in 7 patients (5 DA, 2 DS): 1 perinatally, 2 in the first year, 4 at median age of 6.5 years (range 4–27 years). Biliary atresia was diagnosed perinatally in 13 patients with DA and in 1 at 9 months. Annular pancreas was discovered in 146 patients (15 DA, 113 DS, 18 not specified): 113 perinatally, 4 children, and 28 adults (median age 33 years, range 4–72 years). In the remaining 58 cases, a combination of biliary and pancreatic abnormalities was diagnosed in 41 DA, 16 DS, 1 not specified duodenal obstruction, in 35 patients perinatally, in 4 infants, and in 19 at median age of 3 years (range 1–40 years).
*Conclusions:* Presence of DA/DS should rise the alert for a possible association with biliary/pancreatic abnormalities. Therefore, a careful intraoperative approach should be adopted when repairing DA/DS to avoid damaging biliary and pancreatic ducts. In addition, appropriate imaging looking for biliary/pancreatic anomalies should be considered in symptomatic patients.

## Introduction

Congenital duodenal obstruction is described in 2.5 to 10 per 100,000 live births and it is mostly due to intrinsic duodenal atresia (DA), causing 40–60% of the cases, or stenosis (DS), found in 35–40% of the patients. DA and DS are the consequence of a failed recanalization of the intestinal lumen in early gestation. Concomitant congenital anomalies affect the patients in 45–85% of the cases: congenital heart disease (30%), malrotation (20–30%), genitourinary and renal anomalies (5–15%), esophageal atresia (5–10%), skeletal malformations (5%), and anorectal malformations (5%). Chromosomal abnormalities, expecially trisomy 21, have been commonly identified in these babies [1]. The definitive management of congenital duodenal obstruction is surgical, with the aim to restore gastrointestinal continuity, avoiding damage of the adjacent structures, in particular biliary and pancreatic ducts. Emerging evidence support the use of laparoscopy, however open surgery remains the most adopted technique [2]. Duodenal obstruction and biliary/pancreatic tract anomalies often occur concomitantly. Although predictable, this association is rarely reported and must be considered to avoid major operative complications. Understanding the association between DA/DS and biliary/pancreatic abnormalities is crucial for optimizing surgical strategies. Given that these conditions often

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https://doi.org/10.1016/j.yjpso.2023.100092
Received 31 August 2023; Received in revised form 22 October 2023; Accepted 23 October 2023
Available online 28 October 2023
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co-occur, failure to recognize this association can lead to major post-operative complications, including damage to the biliary and pancreatic ducts or to the development of recurrent pancreatitis which may occur some time after the surgery. All cases of DA and DS associated with biliary and/or pancreatic abnormalities were analyzed, aiming to better understand this association offering a framework for safer and more effective surgical interventions.

## Material and methods

A comprehensive literature search was carried out through PubMed, Science Direct, Scielo, and Scopus, searching the key words "duodenal atresia", "duodenal stenosis", in combination with "biliary malformation", "pancreatic malformation", "annular pancreas", "pancreaticobiliary maljunction". The study was performed according to PRISMA guidelines.

### *Study selection and data extraction*

Three reviewers (FP, SM and CC) screened all titles and abstracts to assess whether each report was potentially eligible for inclusion and whether or not the full text was available. In case of disagreement FP made the final decision. The relevant full text articles were then reviewed and the data extracted.

### *Inclusion and exclusion criteria*

All written studies in English, Spanish, German, Portuguese and Italian reporting any association between DA/DS and biliary and/or pancreatic malformation were included. No limits were set with regard to date of publication. The references of each article were also reviewed to include useful studies that might have been missed with the initial literature screening.

No limits were set on the age of the population.

All study designs were eligible for inclusion. Case reports and case series were also included.

The articles published in a language unknown to the authors and without an English abstract, and those providing insufficient data were excluded. All studies in which the DA/DS was not found in association with biliary and/or pancreatic malformation were excluded as well.

### *Stastistical analysis*

The statistical analysis was performed using GraphPad Prism 6.0. All available data were analyzed and the quantitative data have been presented in a descriptive manner as median and percentage.

## Results

The database search resulted in 522 potentially relevant studies. After the removal of duplicates, 285 studies were identified; 182 studies were excluded for not meeting the inclusion criteria. Finally, 104 studies met the criteria and were used for analysis. A total of 249 patients were considered (Fig. 1): 179 of them were newborns, 7 infants, 25 children, and 38 adults (>15 years).

The list of studies reporting cases of association between DA (Table 1) or DS (Table 2) and biliary and/or pancreatic malformations with clinical data were summarized.

### *Type of biliary or pancreatic abnormalities associated with duodenal atresia or stenosis*

The biliary and pancreatic anomalies associated with DA or DS could be divided into 4 groups:

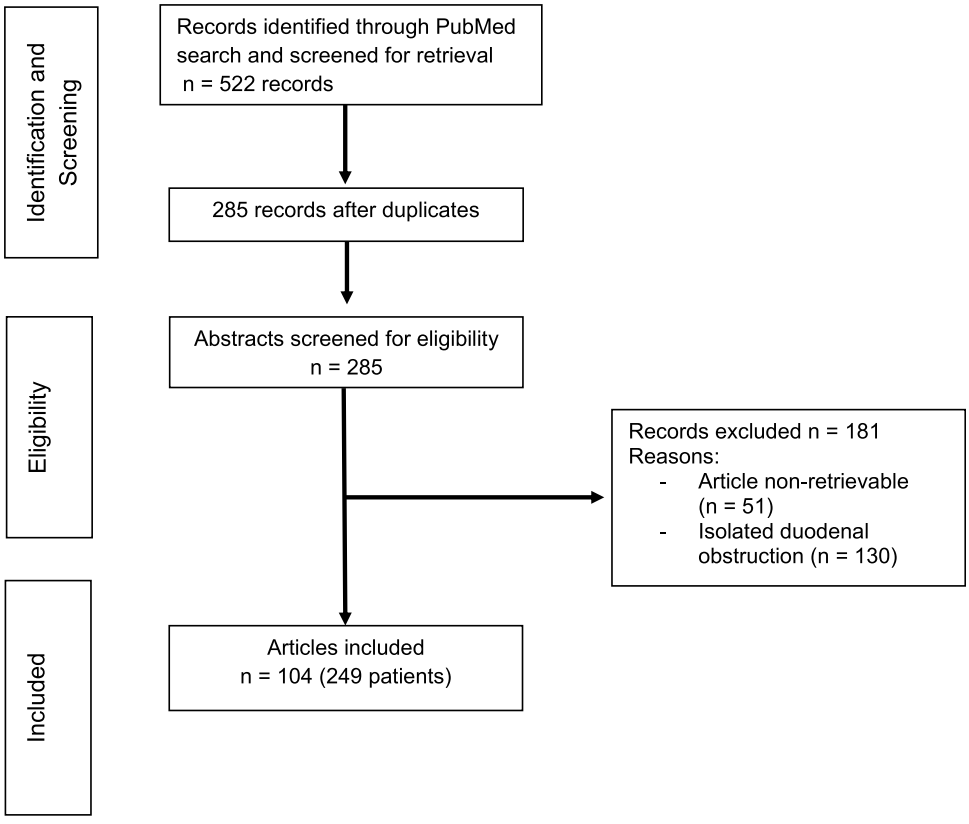



**Fig. 1.** Study selection flowchart.

**Table 1**
List of studies reporting cases of association between duodenal atresia and anomalous bifurcated bile duct, choledocal cyst, biliary atresia, annular pancreas and a combination of biliary and pancreatic abnormalities.

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Karpa, 1906 | 1 nr | no | duodenal atresia | nr | none | no | anomalous bifurcated bile duct conduit between proximal and distal segments at necropsy | nr | none | death at D4 |
| Katz, 1930 | 1 M | no | duodenal atresia | nr | none | no | anomalous bifurcated bile duct conduit between proximal and distal segments at necropsy | nr | none | death at D7 |
| Astley, 1969 | 2<br>1 F | no | duodenal atresia | at term | none | Trisomy 21, ventricular septal defect | anomalous bifurcated bile duct conduit between proximal and distal segments at necropsy | nr | none | death at D18 |
| | 1 F | no | duodenal atresia type I | nr | none | rectoanal atresia, esophageal atresia type III, intestinal malrotation, aorticopulmonary window, left renal agenesis, right hydroureter | anomalous bifurcated bile duct conduit between proximal and distal segments at necropsy | nr | none | death at D2 for subdural hemorrhage |
| Kassner, 1972 | 1 M | no | duodenal atresia | at term | at birth, laparotomy and CDH repair, at D8 re-laparotomy duodenojejunostomy | left CDH, hyperplastic right kidney, accessory adrenal tissue in the mesentery | anomalous bifurcated bile duct conduit between proximal and distal segments. Neither duct terminated in a mucosal papilla. Tile dorsal pancreatic duct joined the ventral pancreatic duct to form the main pancreatic duct, which then entered the longer branch of the common bile duct at right angles. The accessory pancreatic duct was not identified at necropsy | nr | none | death at D1 from sepsis |
| Reid, 1973 | 1 nr | no | duodenal atresia | nr | none | none | anomalous bifurcated bile duct conduit between | nr | none | death at D4 |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | proximal and distal segments at necropsy | | | |
| Lochbuhler, 1988 | 1 nr | nr | duodenal atresia type I | nr | nr | none | anomalous bifurcated bile duct conduit between proximal and distal segments | none | none | nr |
| Knechtle, 1990 | 2<br>1 F | no | duodenal atresia type I | at term | duodenoduodenostomy at D8 | none | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery | none | none | well |
| | 1 F | no | duodenal atresia type III | at term | duodenoduodenostomy at D23 | intestinal malrotation | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery | none | none | well |
| Tashjian, 2001 | 1 M | no | duodenal atresia | 31 | duodeno-duodenostomy | none | anomalous bifurcated bile duct conduit between proximal and distal segments | nr | none | nr |
| Komuro, 2011 | 10<br>1 M | duodenal atresia | duodenal atresia type III | 37 | nr at D2 | PDA, endocardial cushion defect, hydronephrosis, malrotation, vertebra anomaly | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | none | none | death at D3 from cardiac failure |
| | 1 F | no | duodenal atresia type III | 40 | nr at D10 | imperforate anus, ventricular septum defect | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | none | none | well |
| | 1 M | no | duodenal atresia type III | 38 | nr at D10 | none | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | none | none | well |
| | 1 M | no | duodenal atresia type II | 37 | nr at D18 | none | anomalous bifurcated bile duct conduit between | none | none | well |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | | proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | | | |
| | 1 M | no | duodenal atresia type III | 36 | nr at D4 | PDA, endocardial cushion defect | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | none | none | death at D17 from cardiac failure |
| | 1 M | no | duodenal atresia type I | 40 | nr at D4 | malrotation | conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | none | none | well |
| | 1 M | no | duodenal atresia type III | 35 | nr at D16 | tetralogy of Fallot | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | cholestasis | none | well |
| | 1 M | no | duodenal atresia type I | 33 | nr at D5 | double oulet right ventricle, endocardial cushion defect, malrotation | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | none | none | well |
| | 1 M | no | duodenal atresia type III | 37 | nr at D2 | none | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | none | none | well |
| | 1 F | no | duodenal atresia type III | 37 | nr at D1 | ventricular septum defect | anomalous bifurcated bile duct conduit between proximal and distal segments at surgery (distal bowel gas at abdominal x-ray) | cholestasis | none | well |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Komuro, 2000 | 1 M | poly-hydramnios | duodenal atresia | 34 | D4: laparotomy, duodeno-duodenostomy and colostomy for imperforate anus | imperforate anus, cardiovascular anomalies, brain atrophy, tracheobronchial stenosis | choledochal cyst at laparotomy at D4 | at 7 months cholangitis treated by percutaneous transhepatic cholangio-drainage | choledochoduodenostomy at 8 months | death at 8 months from respiratory distress |
| Oowari, 2003 | 1 F | nr | duodenal atresia type III | nr | duodeno-duodenostomy at D1 | no | fusiform dilatation of the common bile duct, fusiform dilatation of the intrahepatic bile duct at 8 years | nr | nr | nr |
| Piessen, 2006 | 1 F | nr | duodenal atresia | nr | operated in childhood for duodenal atresia | no | choledocal cyst in the pancreatic head at 27 years | cholangiolitis | pancreatico-duodenectomy | well |
| Mali, 2007 | 1 M | duodenal atresia | duodenal atresia | 38 | duodeno-duodenostomy in perinatal period | no | choledocal cyst type 1 at 3 months | cholestasis | at 4 months excision of the cyst and anastomosis between common hepatic duct and Roux loop of jejunum. At 6 months cholangitis and revised hepaticojejunostomy for stenosis | well at 2.5 years |
| Goh, 2019 | 1 M | nr | duodenal atresia | 35 | perinatal duodeno-duodenostomy and Ladd's procedure | intestinal malrotation, microtia, bilateral hearing loss | type I choledochal cyst at laparotomy at 7 months | obstructive jaundice and cholangitis at 5 months | nr | nr |
| Reid, 1973 | 1 M | nr | duodenal atresia | nr | duodeno-jejunostomy at D4 | no | biliary atresia at necropsy | hyperbilirubinemia | none | death at D23 |
| Anneren, 1998 | 1 F | duodenal atresia | double duodenal atresia | 36 | double duodenal anastomosis | Cardiac malformation (atrial septum primum), intestinal malrotation | extrahepatic biliary duct atresia, Intrahepatic biliary duct atresia | nr | Kasai portoenterostomy | death at D81 |
| Devine, 2000 | 1 nr | no | duodenal atresia | nr | nr | left isomerism, bilateral left atrial appendages, polysplenia, abdominal heterotaxy | extrahepatic biliary atresia | nr | nr | nr |
| Lawrence, 2000 | 1 nr | nr | duodenal atresia | nr | nr | nr | biliary atresia with pre-pyloric vein | nr | nr | nr |
| Lane, 2000 | 1 M | upper Gi obstruction | distal duodenal atresia | 35 | tapering duodenoplast, duodenal-colonic anstomosis, gastrostomy | severe aplasia cutis congenita, absence of entire midgut | biliary atresia D5 | increased direct bilirubin without bile from gastrostomy | none | death at 3 weeks |
| Deveci, 2000 | 1 M | polyhydramnios | duodenal atresia | 34 | nr | Biliary atresia-splenic malformation sundrome: hypoplasic, unibronchial, unilobed right lung, left aortic arch, patent ductus arteriosus, complex cardiac malformation, esophageal atresia type III, polisplenia, vertebral and rib anomalies | absence of extrahepatic biliary tract | nr | nr | death at 6 h |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Maegawa, 2006 | 1 F | duodenal atresia and IUGR | duodenal atresia | 36 | laparotomy and procedure nr | IUGR, coarse and dysmorphic facial features, microtia with abscence of external auditory meati, fusion of ossicles and Mondini dysplasia, anal malformation, thyroid hemiaplasia, abnormal architecture of the liver vessels (cavernous transformation and portal venous thrombosis), intestinal malrotation, apple peel intestinal atresia | intrahepatic biliary atresia at birth by US scan | nr | nr | death at 5 months |
| Patel, 2014 | 1 M | duodenal atresia and right multicystic kidney | duodenal atresia | 38 | esophageal atresia repair and diamond shaped duodeno-duodenostomy | esophageal atresia type III, right multicystic kidney, hypoplastic left thumb | biliary atresia at D4 | hyperbilirubinemia, pale stools, and dark urine | Kasai portoenterostomy | well at 9 months |
| Kerkeni, 2015 | 1 M | polyhydramnios | duodenal atresia | at term | latero-laterale duodeno-duodenostomy | cardiac malformation, vascular malformation (situs inversus) | biliary atresia at D3 | nr | hepatoportoenterostomy (Kasai) | well at 23 years |
| Durkin, 2017 | 4 nr | nr | duodenal atresia | premature (female 30) | laparotomy | nr | biliary atresia day 4, day 11, day 266 (female) | cholestatic jaundice | Kasai portoenteroanastomosis (3), primary transplant (1) | nr |
| Hays, 1961 | 1 nr | no | duodenal atresia | nr | duodeno-jejunostomy | trisomy 21, intestinal malrotation | annular pancreas at surgery | nr | none | death for sepsis |
| Astley, 1969 | 1 F | no | duodenal atresia | at term | nr | none | annular pancreas at necropsy | nr | none | death at D2 for subdural hemorrhage |
| Clark, 1984 | 1 M | duodenal atresia | duodenal atresia | 33 | duodenoduodenostomy at day 17 | trisomy 21 | annular pancreas at surgery | nr | none | nr |
| Maruiwa, 1988 | 1 F | poly-hydramnios | duodenal atresia | 36 | nr | Cornelia de Lange syndrome, infantile hemangioendothelioma, patency of foramen ovale | annular pancreas at necropsy | nr | none | death at D40 |
| Adeyemi, 1988 | 1 M | no | duodenal atresia | 34 | duodenojejunostomy | no | annular pancreas at surgery | nr | none | nr |
| Baumgartner, 1992 | 1 M | no | duodenal atresia | nr | piloro-duodenostomy at D7 | intestinal malrotation, congenital small glottic region | annular pancreas at surgery | nr | none | nr |
| Weber, 1999 | 1 nr | duodenal atresia | duodenal atresia | nr | duodeno-jejunostomy | trisomy 21, ventricular septal defect, jejunal atresia apple peel | annular pancreas | none | none | nr |
| Fernandez-Gonzalez, 2002 | 1 M | duodenal atresia | duodenal atresia | preterm | nr | Jacobsen syndrome, talipes equinovarus, pancytopenia, | annular pancreas | nr | none | nr |
| Gluer, 2002 | 1 nr | duodenal atresia | duodenal atresia | at term | laparoscopic duodeno-duodenostomy and Ladd's procedure | intestinal malrotation | annular pancreas at surgery | nr | none | nr |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Hsu, 2003 | 1 F | duodenal atresia | duodenal atresia type I | 38 | diamond shaped anastomosis through laparotomy | no | annular pancreas at surgery | none | none | nr |
| Mirza, 2008 | 1 M | no | duodenal atresia | | duodeno-duodenostomy | trisomy 21, intestinal malrotation | annular pancreas at surgery | nr | none | nr |
| Yigiter, 2010 | 22 10 M; 12 F | 12 | 1 duodenal atresia; 1 duodenal web | mean 35 | 9 duodeno-duodenostomy; 9 duodenojejunostomy; 4 gastrojejunostomy | 8 trisomy 21; 1 trisomy 18; 5 malrotation; 4 Meckel diverticulum; 1 Morgagni hernia; 3 anal atresia; 12 cardiovascular anomalies; 1 hypothyroid; 1 cleft limb | 22 annular pancreases at surgery (20 in the first week; 1 on the second week; 1 in the first month) | nr | none | 8 deaths (3 deaths for Candida-related sepsis; 1 death due to renal failure; 1 death due to pneumonia/ septicemia; 1 death during surgery; 1 death at D16) |
| Mirza, 2012 | 1 M | no | duodenal atresia | nr | duodeno-duodenostomy and Ladd's procedure at D3 | intestinal malrotation, trisomy 21 | annular pancreas at D3 | nr | none | death at D16 for sepsis |
| Balakumar, 2014 | 1 F | no | duodenal atresia | nr | diamond shaped duodeno-duodenostomy | trisomy 21, coarctation of the aorta | annular pancreas at surgery | none | none | well at 20 months |
| Jadhav, 2021 | 1 F | no | duodenal atresia | term | esophageal atresia repair at birth At D9 gastro-jejunostomy and Ladd's procedure, partial right lobectomy | esophageal atresia type III, intestinal malrotation, communicating bronchopulmonary foregut malformation | annular pancreas at surgery | none | none | well at 6 months |
| Reid, 1973 | 7 1 nr | nr | duodenal atresia | nr | nr | tracheoesophageal fistula, intestinal malrotation, partial situs inversus, imperforate anus, cardiovascular anomalies, urogenital anomalies, arthrogryposis | annular pancreas, non-communicating cyst duct, stenotic common bile duct at necropsy | none | none | death at D1 |
| | 1 nr | no | duodenal atresia | nr | duodeno-duodenostomy at D3 | no | non-communicating common bile duct, pancreatic ductal obstruction, obstructive biliary cirrhosis at necropsy | none | none | death at D11 |
| | 1 F | no | duodenal atresia | nr | nr | trisomy 21 | no ampulla of Vater, non-communicating cystic duct | none | none | nr |
| | 1 F | no | duodenal atresia type I (web) | nr | excision of the web | no | no ampulla of Vater at surgery | hyperbilirubinemia | none | nr |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | 1 nr | no | duodenal atresia | nr | esophageal fistula divided, gastrostomy, duodenojejunostomy | esophageal atresia, atrial septal defect | absence of gallbladder at surgery | none | none | nr |
| | 1 M | no | duodenal atresia | nr | nr | esophageal atresia, anorectal malformation, bilateral talipes, hypoplastic lungs, absent right ureter and kidney, multicystic left kidney, bifid scrotum, congenital dislocation of the hips, two umbilical arteries, ectopic pancreas | absence of gallbladder and cystic duct | none | none | nr |
| | 1 M | no | duodenal atresia | 34 | duodeno-duodenostomy and gastrostomy at day 5 | Meckel's diverticulum, small diverticulum on the antimesenteric border of the jejunum | abscence of gallbladder and cystic duct at surgery | none | none | nr |
| Patti,1985 | 1 F | no | duodenal atresia | nr | esofagostomy and gastrostomy, transmesocolic isoperistaltic duodenojejunal anastomosis at birth. At 8 months retrosternal esophago-colon-gastroplasty | esophageal atresia type I | preduodenal positions of the common bile duct and portal vein at first surgery | none | none | well at 12 months |
| Heij 1987 | 1 M | no | duodenal atresia | term | side to side duodeno-duodenostomy | no | annular pancreas, agenesis of the gallbladder, dilation of the common bile duct at first surgery: observation and cholangiogram | jaundice | none | well |
| Martinez-Frias, 1992 | 2 siblings 1 F | no | duodenal atresia | at term | none | dysmorphic face | Extrahepatic biliary atresia, hypoplastic pancreas at necropsy | nr | none | death at D2 |
| | 1 M | no | duodenal atresia | at term | none | esophageal atresia type III, hypospadias, hypoplastic intestine | hypoplastic pancreas and gallbladder at necropsy | nr | none | death at D5 |
| Coughlin, 1992 | 2 1 M | duodenal atresia | duodenal atresia | nr | duodeno-duodenostomy and colostomy | imperforate anus, hypospadias, ventricular septal defect, patent ductus arteriosus, left hydronephrosis | absence of gallbladder at surgery | none | none | well at 6 months |
| | 1 M | duodenal atresia | duodenal atresia | nr | duodeno-duodenostomy | no | absence of gallbladder at surgery | none | none | well at 5 months |
| Okada, 1993 | 2 1 M | nr | duodenal atresia type III | 34 | diamond shaped anastomosis through laparotomy at D2 | no | annular pancreas, fusiform dilatation of the common bile | none | portal jejunostomy (roux en Y anastomosis) | nr |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | 1 F | nr | duodenal atresia type III | 40 | diamond shaped anastomosis through laparotomy at D7 | jejunal aberramt pancreas | duct, fusiform dilatation of the intrahepatic bile duct at 12 years annular pancreas, fusiform dilatation of the common bile duct, fusiform dilatation of the intrahepatic bile duct at 3 years | nr | portal jejunostomy (roux en Y anastomosis) | nr |
| Anneren, 1998 | 2 sibs, both in vitro M | duodenal atresia | double duodenal atresia | 40 | double duodenal anastomosis | esophagela atresia type III, intestinal malrotation | Extrahepatic biliary duct atresia, Intrahepatic biliary duct atresia, hypoplastic pancreas | nr | none | death |
| Pameijer, 2000 | 1 M | duodenal atresia, esophageal atresia type I, trisomy 21 | duodenal atresia | 35 | at birth, eosphageal anastomosis, duodenoduodenostomy with tapering, gastro-jejunostomy | globular annular pancreas, extrahepatic biliary ducts patent but stretched, elongated and disappeared within the pancreas | type I choledochal cyst, biliary and pancreatic ductal atresia (post mortem) at D14 | respiratory difficulty | choledochoduodenostomy | intermittent tolerance of enteral feeding, abdominal sepsis, enterocutaneous fistula, entero-enteral fistula, death at 4,5 months |
| Sugimoto, 2002 | 1 F | polyhydramnios | duodenal atresia (Vater's ampulla distal) | 38 | diamond shaped anastomosis through laparotomy | no | choledocal cyst and pancreaticobiliary maljunction at 32 months | abdominal pain, jaundice | hepaticoduodenostomy | well |
| Mitchell, 2004 | 3<br>1 M | duodenal atresia | duodenal and jejunal atresia | 36 | at D3 laparotomy and atresias repair | neonatal diabetes, RFX6 mutation | hypoplastic pancreas, intrahepatic biliary atresia, and absence of gallbladder at surgery D3 | cholestasis | none | death at 5 months, multiple organ failure |
| | 1 F | duodenal atresia | duodenal and jejunal atresia (type IIIA) | 34 | at day 1 laparotomy and atresias repair | neonatal diabetes, RFX6 mutation | annular pancreas, intrahepatic biliary atresia, and absence of gallbladder at surgery D1 | cholestasis | none | death at 6 months, multiorgan failure |
| | 1 M | duodenal atresia | duodenal and jejunal atresia | 38 | in the first week laparotomy and atresias repair | neonatal diabetes, RFX6 mutation | hypoplastic pancreas, intrahepatic biliary atresia and hypoplastic gallbladder at surgery | conjugated hyperbilirubinemia, colorless stools | none | death at 4 months |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Shih, 2005 | 1 F | polyhydramnios | duodenal atresia type III | 41 | duodeno-duodenostomy at D3 | no | annular pancreas, Alonso Lej type IV choledochal cyst with pancreaticobiliary maljunction at 7 years | abnormal liver and pancreatic function | cholecystectomy and hepaticoduodenostomy | well |
| Mali, 2007 | 2<br>1 M | duodenal atresia | duodenal atresia | 28 | on day 3 duodeno-duodenostomy | no | cyst in the confluence of two right hepatic duct, a single left hepatic duct and cystic duct suspected at 3 weeks | at 3 months jaundice, failure to thrive, non-bilious vomiting | Roux en Y jejunal loop anastomosed to all 3 hepatic ducts | well |
| | 1 M | polyhydramnios | double duodenal atresia | 33 | duodenojejunostomy, Ladd's procedure | intestinal malrotation | dilated common bile duct at 14 months | jaundice, conjugated hyperbilirubinemia | duodeno-duodenostomy (distal to the duodenojejunostomy) + excision of the extrahepatic bile ducts and Roux en Y hepaticojejunostomy | well |
| Galan-Gomez, 2007 | 1 F | duodenal atresia | duodenal atresia | 38 | duodeno-duodenostomy and Ladd's procedure | intestinal malrotation, | hypoplastic pancreas and agenesis of extrahepatic biliary ducts and gallbladderat birth | cholestasis | none | death at D60 |
| Chappel, 2008 | 1 F | duodenal atresia | duodenal atresia | 35 | at day 1 laparotomy, duodeno-duoenostomy, Ladd's procedure | intestinal malrotation, anorectal malformation, neonatal diabetes, RFX6 mutation | gallbladder agenesis | conjugated hyperbilirubinemia | none | well 6 years |
| Okuyama, 2008 | 1 M | no | duodenal atresia | nr | duodeno-duodenostomy | no | annular pancreas, pancreas divisum and pancreaticobiliary maljunction at 2,5 years | recurrent pancreatitis | Frey procedure (local resection of the head of pancreas in combination with longitudinal pancreaticojejunostomy) and roux-en-y pancreatico- and hepatico-jejunostomy | 20 months after ok |
| Iwai, 2009 | 1 F | duodenal atresia | duodenal atresia type III | 37+3 | duodeno-duodenostomy | no | annular pancreas, fusiform type choledochal cyst with pancreatobiliary maljunction at 4 years | cholangitis and pancreatitis | excision of choledochal cyst and hepaticojejunostomy | NR |
| Martinovici 2010 | 1 M | duodenal atresia | duodenal atresia | 38 | at D2 duodeno-jejunostomy | apple peel-type jejunal atresia, intestinal malrotation, neonatal diabetes, IUGR, hemochromatosis, RFX6 mutation | gallbladder agenesis and biliary atresia, pancreas hypoplasia at D2 | conjugated hyperbilirubinemia | none | death at 2 months |

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**Table 1** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Urushihara, 2010 | 3<br>1 M | nr | duodenal atresia | nr | at D12 duodeno-duodenostomy | no | annular pancreas, pancreatobiliary malunion at 4 years | 1-year history of recurrent pancreatitis | Excision of extrahepatic bile duct, hepaticojejunostomy | well |
| | 1 F | nr | duodenal atresia | nr | at D2 gastro-jejunostomy | Trisomy 21, ventricular septal defect | annular pancreas, pancreatobiliary malunion, intraluminal duodenal diverticulum at 21 months | 6-months history of recurrent pancreatitis | Excision of extrahepatic bile duct, hepaticojejunostomy with duodenojejunostomy, marsupialization of intraluminal duodenal diverticulum | well |
| | 1 F | nr | duodenal atresia | nr | at D0 duodeno-duodenostomy | no | annular pancreas, choledochocele at 11 years | 2-years history of recurrent pancreatitis | Marsupialization of choledochocele, plasty of pancreatic duct orifice | well |
| Spiegel, 2011 | 1 M | duodenal atresia | duodenal atresia | 38 | duodeno-duodenostomy | neonatal diabetes, jenjunal atresia | annular pancreas and gallbladder agenesis at 1 month | cholestatic jaundice | none | well at 21 months |
| Komuro, 2011 | 1 M | yes: duodenal atresia | duodenal atresia type III | 37 | nr at D2 | PDA, endocardial cushion defect, hydronephrosis, malrotation, vertebra anomaly | pancreatic duct from the accessory pancreatic duct to the main ampulla at surgery | nr | none | death at D3 for cardiac failure |
| Concepcion, 2014 | 1 M | duodenal atresia, IUGR | duodenal atresia | 34 | at D5 laparotomy, malrotation correction and Roux-en-Y anastomosis | malrotation, mutation in transcription factor RFX6, neonatal diabetes | annular pancreas, gallbladder agenesis at 3 months | cholestasis, diarrhea | none | liver failure and death at 5 months |
| Zoetsch, 2015 | 1 F | duodenal atresia | duodenal atresia | 38 | duodeno-duodenostomy | no | intrapancreatic choledochocele type IIIat 3 months | hyperbilirubinemia and GGT elevated | transduodenal marsupialization | nr |
| Kulkarni, 2017 | 1 M | duodenal atresia and no gallbladder | duodenal atresia | at term | nr | PDX1 gene mutation, neonatal diabetes | annular pancreas, hypoplastic gallbladder | nr | none | well at 2 years |
| Puvabanitsin, 2018 | 1 F | no | duodenal atresia | 39 | nr | malrotation, trisomy 21 | anomalous common bile duct at birth | none | none | nr |
| Yokoyama, 2020 | 1 F | no | duodenal atresia type III | 36+6 | diamond shaped anastomosis through laparotomy | 1p36 deletion syndrome (intellectual disability) | Pancreaticobiliary maljunction (pancreatic and bile ducts join outside the duodenal wall), Vater ampulla in distal duodenum, no annular pancreas 39 months | fever, abdominal pain, acholic stool and jaundice; high serum biliary enzyme levels | Extrahepatic bile duct resection from the porta hepatis to the level of confluence of the main pancreatic duct, and hepaticojejunostomy | nr |
| Downing, 2020 | 1 F | duodenal atresia | duodenal atresia | 32 | duodeno-duodenostomy and Ladd's procedure at D4 | intestinal malrotation | choledochal cyst, annular pancreas at 2 years choledochal cyst | feeding intolerance, jaundice | cholecystectomy, choledochal cyst excisione and hepatico-duodenostomy | well at 3 years |

**Table 2**
List of studies reporting cases of association between duodenal stenosis and anomalous bifurcated bile duct, choledocal cyst, biliary atresia, annular pancreas and a combination of biliary and pancreatic abnormalities.

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Raine, 1977 | 1 M | no | duodenal stenosis | 37 | duodeno-duodenostomy | right heart hypoplasia, pulmonary stenosis, ventricular septal defect | anomalous bifurcated bile duct conduit between proximal and distal segments, bilobed gallbladder at laparotomy | none | none | well |
| Komuro, 2011 | 3<br>1 M | duodenal obstruction | duodenal stenosis | 38 | nr at birth | intestinal malrotation | anomalous bifurcated bile duct conduit between proximal and distal segments, annular pancreas at birth | none | none | well |
| | 1 M | no | duodenal stenosis | 38 | nr at D4 | none | anomalous bifurcated bile duct conduit between proximal and distal segments, annular pancreas at birth | none | none | well |
| | 1 M | no | duodenal stenosis | 39 | nr at D4 | patent ductus arteriosus | anomalous bifurcated bile duct conduit between proximal and distal segments, annular pancreas | none | none | well |
| Nakamura, 1993 | 1 F | nr | duodenal stenosis | 37 | nr | trisomy 21, ventricular septal defect | fusiform dilatation of the common bile duct at 5 years | nr | portal jejunostomy (roux en Y anastomosis) | nr |
| Mali, 2007 | 1 M | no | duodenal stenosis diagnosed at 10 years | nr | duodeno-duodenostomy at 10 years | superior mesenteric artery (SMA) syndrome | choledochal cyst at 6 years | nr | excision of choledocal cyst and hepaticojejunostomy at 6 years | nr |
| Jolley, 1992 | 1 F | no | duodenal web | 36 | side-to-side duodenojejunostomy at the time of biliary atresia correction | apple peel-type jejunal atresia, intestinal malrotation | biliary atresia at birth | nr | portoenteroanastomosis with Roux en Y using apple peel jejunum as the Roux, side-to-side duodeno-jejunostomy and Ladd's procedure | well at 4 years |
| Gross, 1944 | 1 M | no | duodenal stenosis diagnosed at D3 | term | duodeno-jejunostomy at D3 | intestinal malrotation | annular pancreas at surgery | none | none | nr |
| Lehman, 1942 | 1 M | no | duodenal stenosis diagnosed at 23 years | nr | none | no | annular pancreas at 23 years | abdominal pain | division of annular pancreas | nr |
| Wilson, 1953 | 3<br>1 M | no | duodenal stenosis | nr | gastro-jejunostomy | no | annular pancreas at D18 | nr | none | well |
| | 1 M | no | duodenal stenosis | nr | duodeno-jejunostomy | no | annular pancreas at 20 months | nr | none | well |

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**Table 2** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | diagnosed at 20 months | | | | | | | |
| | 1 M | no | duodenal stenosis diagnosed at D7 | nr | duodeno-jejunostomy | no | annular pancreas at D7 | nr | none | death at D 10 for aspiration pneumonia |
| Mast, 1957 | 8<br>1 M | no | duodenal stenosis diagnosed at 66 years | nr | none | no | annular pancreas at 66 years | symptoms of obstruction at 24 years | resection of annular pancreas: bad idea; then vagotomy | nr |
| | 1 M | no | duodenal stenosis diagnosed at 24 years | nr | nr | no | annular pancreasat 24 years | symptoms of obstruction | nr | nr |
| | 1 M | no | duodenal stenosis at 59 years | nr | gastro-enterostomy | no | annular pancreas at 59 years | abdominal pain for 35 years | none | death at 61 years for obstructive jaundice and acute pancreatitis |
| | 1 M | no | duodenal stenosis at 57 years | nr | gastro-enterostomy and vagotomy | no | annular pancreas at 57 years | abdominal pain at 57 years | none | nr |
| | 1 M | no | duodenal stenosis diagnosed at 34 years | nr | gastro-enterostomy | no | annular pancreas at 34 years | symptoms of obstruction at 34 years | none | nr |
| | 1 F | no | duodenal stenosis diagnosed at 30 years | nr | gastro-enterostomy | no | annular pancreas at 30 years | symptoms of obstruction, abdominal pain, vomiting | none | nr |
| | 1 F | no | duodenal stenosis diagnosed at 39 years | nr | nr | no | annular pancreas at 39 years | symptoms of obstruction, abdominal pain, vomiting | nr | nr |
| | 1 M | no | duodenal stenosis diagnosed at 5 weeks | nr | duodeno-jejunostomy | patent foramen ovale and ductus arteriosum | annular pancreas at 5 weeks | vomiting and jaundice | none | death 2 days after for pneumonia |
| Hays, 1961 | 6<br>1 nr | no | duodenal stenosis | term | duodeno-jejunostomy | no | annular pancreas at surgery | nr | none | nr |
| | 1 nr | poly-hydramnios | duodenal stenosis | nr | duodeno-jejunostomy | no | annular pancreas at surgery | nr | none | nr |
| | 1 M | poly-hydramnios | duodenal stenosis | nr | duodeno-jejunostomy | no | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis at D6 | nr | duodeno-jejunostomy | no | annular pancreas at D6 | nr | none | nr |
| | 1 M | poly-hydramnios | duodenal stenosis | nr | gastrostomy and colostomy | esophageal atresia type III, imperforate anus | annular pancreas at surgery | nr | none | death of respiratory distress 3 days after surgery |
| | 1 nr | poly-hydramnios | duodenal stenosis | nr | esophageal atresia repair and duodeno-jejunostomy | esophageal atresia type III | annular pancreas at surgery | nr | none | death 3 days after surgery for re-fistula |
| Merrill, 1976 | 24<br>1 F | no | duodenal stenosis | nr | duodeno-jejunostomy | trisomy 21 | annular pancreas at surgery | nr | none | |

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**Table 2** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | 1 F | no | duodenal stenosis | nr | colostomy | malrotation, coartation, AD, VSD, PDA, persistent left vena cava | annular pancreas at surgery | nr | none | death of aspiration pneumonia |
| | 1 M | no | duodenal stenosis | nr | duoden-ojejunostomy and colostomy | malrotation, imperforate anus, sacral agenesis | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis | nr | duodeno-jejunostomy, duodeno-duodenostomy | no | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | duodeno-duodenostomy | no | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | duodeno-duodenostom, Meckel's resection, aortopulmonary artery shunt | malrotation, Meckel's diverticulum, tetralogy, absent left radius | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | duodeno-jejunostomy, aortopulmonary artery shunt | tetralogy | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | gastrostomy, duodeno-jejunostomy, ligation TEF, colostomy | esophageal atresia type III, imperforate anus, CHD, double uterus | annular pancreas at surgery | nr | none | death of cardiac arrest |
| | 1 M | no | duodenal stenosis | nr | duodeno-jejunostomy | trisomy 21, tetralogy, ectopic pancreatic tissue | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | duodeno-jejunostomy | trisomy 21 | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | duodeno-jejunostomy | no | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | duodeno-duodenostomy | no | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | duodeno-duodenostom, ligation TEF, gastrostomy | trisomy 21, esophageal atresia type III, malrotation, Meckel's diverticulum | annular pancreas at surgery | nr | none | death of bronchopneumonia |
| | 1 F | no | duodenal stenosis | nr | duodeno-jejunostomy | Trisomy 21, Meckel's diverticulum | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis | nr | duodeno-jejunostomy and pull through | Imperforate anus | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis | nr | duodeno-jejunostomy | Trisomy 21 | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis | nr | pull through, gastro-duodenostomy, gastro-jejunostomy | Trisomy 21, imperforate anus | annular pancreas at surgery | nr | none | death of aspiration pneumonia |
| | 1 F | no | duodenal stenosis | nr | duodeno-duodenostomy | none | annular pancreas at surgery | nr | none | nr |

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**Table 2** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | 1 M | no | duodenal stenosis | nr | duodeno-jejunostomy | intestinal malrotation, rectal stricture, multiple congenital anomalies | annular pancreas at surgery | nr | none | death of bronchopneumonia |
| | 1 M | no | duodenal stenosis | nr | gastrostomy, esophageal atresia repair, duodeno-duodenostomy | esophageal atresia type III | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis | nr | gastrostomy, esophageal atresia repair, duodeno-jejunostomy | esophageal atresia type III | annular pancreas at surgery | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | none | multiple central nervous system anomalies | annular pancreas at necropsy | nr | none | death |
| | 1 F | no | duodenal stenosis and preduodemal portal vein at 3 weeks of age | nr | release of midgut volvolus | intestinal malrotation, preduodenal portal vein | annular pancreas at 3 weeks of age | nr | none | nr |
| | 1 F | no | duodenal stenosis diagnosed at 11 years | nr | duodeno-jejunostomy at 11 years | none | annular pancreas at 11 years | nr | none | nr |
| Kiernan, 1980 | 15 (6 neonates [4 M and 2 F], 9 adults [5 M and 4F] | nr | duodenal obstruction | nr | 1 neonate gastrojejunostomy; 5 neonates duodeno-duodenostomy. 1 adult truncal vagotomy, 60% gastrectomy and gastroduodenal reconstruction, but annular pancreas was diagnosed after obstruction and gastrojejunostomy was performed; 1 adult with duodenal ulcer 70% gastrectomy and gastrojejunal reconstruction; 4 adults duodenoduodenostomy; 2 adults posterior gastrojejunostomy; 1 adult excision anterior part of pancreas annulus and died | 1 esophageal atresia type III | annular pancreas | nr | none | 2 neonates died of pnuemonia (1 with AE type III and 1 with gastrojejunostomy) |
| Tchirkow, 1980 | 1 F | no | duodenal stenosis | nr | duodeno-jejunostomy at D9 cholecystectomy at 9 years | no | annular pancreas at perinatal surgery | cholecystitis with hydrops at 9 years | none | nr |
| Brambs, 1986 | 1 F | no | duodenal stenosis diagnosed at 28 years | nr | none | no | annular pancreas at 28 years | colicky pain | none | nr |
| Adeyemi, 1988 | 2 1 F | no | duodenal stenosis | term | excision of the diaphragm | no | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis at D12 | nr | excision of the diaphragm | partial situs inversus | annular pancreas at surgery | nr | none | nr |
| Synn, 1988 | 3 nr | nr | duodenal atresia or stenosis | nr | 5 duodenoduodenostomy; 8 duodenojejunostomy; 2 gastroenterostomy | 4 trisomy 21; 3 cardiac anomalies; 2 malrotations | annular pancreas at surgery | nr | none | nr |

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**Table 2** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Bickler, 1992 | 1 nr | nr | duodenal web | 37 | nr | no | annular pancreas at surgery | nr | none | nr |
| Kelly, 1993 | 1 M | no | duodenal stenosis | nr | pancreatoduodenectomy, distal gastrectomy, cholecystectomy | no | annular pancreas at 43 years | epigastric pain, vomiting, weight loss gastric carcinoma | none | well after 11 years |
| Balustein, 1993 | 1 M | no | duodenal stenosis diagnosed at 10 months | 36 | duodenoduodenostomy | Rothmund-Thomson syndrome | annular pancreas at 10 months | nr | none | nr |
| Olson, 1994 | 1 M | no | duodenal stenosis at 8 months | nr | nr | no | annular pancreas at 8 months | nr | none | nr |
| Hamm, 1997 | 1 M | no | duodenal stenosis diagnosed at 52 years | nr | duodenohemipancreatectomy (Whipple's procedure) | no | annular pancreas at 52 years | nr | none | nr |
| Summer, 1997 | 1 F | no | duodenal stenosis | nr | esophageal atresia repair, duodenoduodenostomy and colostomy at 4 days | esophageal atresia type III, vertebral anomalies, imperforate anus, ambiguous genitalia | annular pancreas at D4 | nr | none | nr |
| Jadvar, 1999 | 7<br>1 M | no | duodenal stenosis | nr | nr | no | annular pancreas at 21 years | abdominal pain and vomiting (symptoms of duodenal obstruction) | none | nr |
| | 1 F | no | duodenal stenosis | nr | nr | no | annular pancreas at 32 years | vomiting (symptoms of obstruction) | none | nr |
| | 1 M | no | duodenal web | nr | nr | no | annular pancreas at 70 years | obstruction | none | nr |
| | 1 M | no | duodenal stenosis | nr | nr | no | annular pancreas at 72 years | vomiting (symptoms of obstruction) | none | nr |
| | 1 M | no | duodenal stenosis | nr | nr | no | annular pancreas at 14 years for trauma during basketball game | nr | none | nr |
| | 1 F | no | duodenal stenosis | nr | nr | no | annular pancreas, dilatation of common bile duct and pancreatic duct at 38 years | recurrent vomiting | none | nr |
| | 1 M | no | duodenal stenosis | nr | nr | no | annular pancreas at 30 years | abdominal pain | none | nr |
| Sencan, 2002 | 2<br>1 F | no | duodenal stenosis | 36 | duodeno-duodenostomy | ventricular septal defect, intestinal malrotation, | annular pancreas at surgery | nr | none | nr |
| | 1 M | no | duodenal stenosis | 40 | duodeno-duodenostomy | trisomy 21, intestinal malrotation | annular pancreas at surgery | nr | none | nr |
| McCollum, 2002 | 1 M | no | duodenal stenosis | nr | duodeno-duodenostomy | no | annular pancreas at 4 years | nr | none | nr |

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**Table 2** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | diagnosed at 4 years | | | | | | | |
| Chirdan, 2004 | 1 nr | no | duodenal stenosis at day 6 | | duodeno-jejunostomy and colostomy | anorectal malformation | annular pancreas at D6 | none | none | nr |
| Miyazawa, 2004 | 1 M | no | duodenal stenosis at 40 years | nr | duodeno-duodenostomy at 40 years | no | annular pancreas at 40 years | nr | none | well |
| Jimenez, 2004 | 16<br>11 M and 5 F | 5 duodenal obstructions | duodenal stenosis diagnosed in 12 pts in the first week, in 1 in the first month and 1 in the first year | 36.6 + 2.8 | 14 duodeno-duodenostomy; 1 duodeno-jejunostomy; 1 Ladd's procedure | 7 malrotations, 2 esophageal atresia type II (1 of them with imperforate anus and trisomy 18), 1 ileal atresia, 1 jejunal ectopic gastric mucosa, 1 hydrocephalus, 1 bilateral hydronephrosis | annular pancreas at surgery | none | none | nr |
| De Ugarte, 2006 | 2<br>1 F | no | duodenal stenosis at 32 years of age | nr | gastro-jejunostomy | no | annular pancreas at 32 years | nr | none | well |
| | 1 M | no | duodenal stenosis diagnosed at 56 years | nr | gastro-jejunostomy | no | annular pancreas at 56 years | nr | none | well |
| Savino, 2007 | 1 F | no | duodenal stenosis diagnosed at 15 years | nr | Roux-en Y duodeno-jejunostomy | trisomy 21 | annular pancreas at 15 years | nr | none | well |
| Ohno, 2008 | 1 M | no | duodenal stenosis diagnosed at 6 years | nr | | no | annular pancreas at 6 years | pancreatitis | none | well |
| Chinnappan, 2008 | 1 M | no | duodenal stenosis diagnosed at 55 years | nr | duodeno-duodenostomy | no | annular pancreas at 55 years | weight loss, anemia, duodenal ulcer | none | nr |
| Yigiter, 2010 | 22<br>10 M; 12 F | 12 | 1 duodenal atresia; 1 duodenal web | mean 35 | 9 duodeno-duodenostomy; 9 duodenojejunostomy; 4 gastrojejunostomy | 8 trisomy 21; 1 trisomy 18; 5 malrotation; 4 Meckel diverticulum; 1 Morgagni hernia; 3 anal atresia; 12 cardiovascular anomalies; 1 hypothyroid; 1 cleft limb | 22 annular pancreases at surgery (20 in the first week; 1 on the second week; 1 in the first month) | nr | none | 8 deaths (3 deaths for Candida-related sepsis; 1 death due to renal failure; 1 death due to pneumonia/ septicemia; 1 death during surgery; 1 death at D16) |
| Mahdi, 2011 | 1 M | no | duodenal stenosis | nr | gastro-enterostomy at 24 years | no | annular pancreas at 24 years | nr | none | well |

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**Table 2** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | diagnosed at 24 years | | | | | | | |
| Markljung, 2012 | 1 M (his mother had the same condition) | no | duodenal web | 39 | duodeno-duodenostomy | no | annular pancreas at surgery | none | none | nr |
| Miranda, 2013 | 1 F | duodenal obstruction | duodenal stenosis | 36 | duodeno-duodenostomy | intestinal malrotation, alveolar capillary dysplasia with misalignment of pulmonary veins | annular pancreas at surgery | none | none | death at D15 |
| Kobayashi, 2016 | 1 F | no | duodenal stenosis diagnosed at 56 years | nr | | no | annular pancreas at 56 years | epigastric and back pain | none | nr |
| Cai, 2018 | 1 M | duodenal obstruction | duodenal stenosis | 39 | duodeno-duodenostomy | Meckel's diverticulum | annular pancreas at surgery | none | none | nr |
| Ladipo-Ajayi, 2020 | 1 F | duodenal atresia | duodenal stenosis | 34 | duodeno-duodenostomy at D4 | dysmorphic face, trisomy 21 | annular pancreas at D4 | pancreatitis | none | death for cardiac arrest at 4 days |
| Jona, 1976 | 1 F | no | duodenal stenosis | nr | excision of the web | no | Vater ampulla in the medial wall of the duodenal web at surgery | none | none | nr |
| Komura, 1993 | 1 F | no | duodenal stenosis diagnosed at 2 years of age | nr | diamond shaped anastomosis through laparotomy at 2 years of age | none | pancreaticobiliary maljunction, annular pancreas at 2 years | nr | portal jejunostomy (roux en Y anastomosis) together with duodeno-duodenostomy | discharged at D13 |
| Gentile, 1999 | 1 M | duodenal atresia | duodenal stenosis | 34 | at 9 days duodeno-duodenostomy, colostomy, malrotation correction | esophageal atresia type III, intestinal malrotation, rectoanal atresia, atrial septal defect | annular pancreas, gallbladder, and extrahepatic biliary duct agenesis at D9 | | Kasai portoenteroanastomosis | death at 10 months of respiratory failure (recurrent bronchiolitis) |
| Komuro, 2000 | 2 M | nr | duodenal stenosis | 29 | at 11 months, side to side duodeno-duodenostomy | no | intrapancreatic cyst + annular pancreas + anomalous pancreatobiliary duct junction at 11 months | hyperamylasemia in the bile | at 13 months, cholecistectomy, resection of common bile duct proximal to the cyst, excision of the cyst, Roux-en-Y choledocojejunostomy | pancreatic fistula subsided spontaneously Healthy at 9 years. |
| Mitchell, 2004 | 1 F | duodenal atresia | duodenal web | 39 | at D2 laparoscopy | malrotation, neonatal diabetes, RFX6 mutation | hypoplastic pancreas and absence of gallbladder at surgery | hyperbilirubinemia, acholic stools | | well |
| Muraoka, 2005 | 1 M | duodenal obstruction | duodenal stenosis | 38 | duodeno-duodenostomy | no | common bile duct ran into ventral wall of duodenum | no | none | nr |

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**Table 2** (*continued*)

| First author and year | Patients n and gender | Prenatal diagnosis | Type of duodenal malformation | Weeks at delivery | Surgical procedure | Other abnormalities | Type and time of diagnosis of biliary/ pancreatic malformation | Symtoms of biliary malformation | Surgical procedure for biliary malformation | Outcome and follow up |
|---|---|---|---|---|---|---|---|---|---|---|
| Urushihara, 2010 | 3<br>1 M | nr | duodenal stenosis | nr | at D3 duodeno-duodenostomy | no | Pancreas divisum at 6 years | 3-years history of recurrent pancreatitis | Pancreaticojejunostomy, cholecystectomy, plasty of pancreatic duct orifice | well |
| | 1 M | nr | duodenal stenosis | nr | at D3 duodeno-duodenostomy | Trisomy 21, ventricular septal defect; hydrocephalus | Choledochocele at 10 years | 3-years history of recurrent pancreatitis | None for familiar refusal | nr |
| | 1 M | nr | duodenal stenosis | nr | at D6 duodeno-duodenostomy | no | Pancreas divisum at 10 years | 6-months history of recurrent pancreatitis | Pancreaticojejunostomy cholecystectomy | well |
| Komuro, 2012 | 2 M | no | duodenal stenosis diagnosed at 3 years of age on upper GI contrast | 34 | duodeno-duodenostomy | trisomy 21, endocardial cushion defect corrected at 1 year of age | annular pancreas + long dorsal pancreatic duct passing anteriorly to encircle the duodenum 3 years | abdominal pain, hyperamylasemia, hyperlipasemia, duodenopancreatic reflux through the ampulla | none | well |
| Cheng, 2013 | 1 M | no | duodenal stenosis diagnosed at 26 years of age | 40 | duodenojejunostomy (side to side) and choledoch-ojejunostomy (Roux-en-Y anastomosis) | no | annular pancreas, pancreatobiliary maljunction (dilatated common bile duct and high confluence of pancreatobiliary ducts)<br>26 years | in the past 23 years 3 episodes of acute abdomen diagnosed as acute pancreatitis, abdominal pain, abnormal liver and pancreatic function, increased levels of plasma amylase | choledocho-jejunostomy (Roux-en-Y anastomosis) | nr |
| Trandafir, 2014 | 1 F | no | duodenal stenosis | 36 | diamond shaped anastomosis through laparotomy and removal of ectopic jejunal pancreas with jejuno-jejunostomy at D27 | trisomy 21 | islet of type 1 ectopic pancreas located in the jejunum at surgery | none | removal of ectopic pancreas | nr |
| Chowdhury, 2018 | 1 F | no | duodenal stenosis diagnosed at 43 years | nr | gastrojejunostomy | no | annular pancreas and dilated common bile duct at 43 years | biliary and pancreatic symptoms | hepaticojejunostomy (Roux-en-y) | well |
| Griggs, 2020 | 1 F | duodenal obstruction | duodenal stenosis | 38 | duodeno-duodenostomy | no | bifid common bile duct and annular pancreas at surgery | none | none | nr |

- anomalous bifurcated bile duct conduit between proximal and distal atretic segments
- choledochal cyst
- biliary atresia
- annular pancreas

Some patients presented a combination of biliary and pancreatic abnormalities.

*Anomalous bifurcated bile duct*

Anomalous bifurcated bile duct was described in 24 (9.6%) cases (6 female, 15 male, 3 not reported), 20 associated with DA, and 4 with DS [3–9]. The diagnosis was reached in 6 patients at necropsy and in 18 intraoperatively at the time of perinatal surgery for the duodenal obstruction. Fourteen patients had other associated congenital malformations. Only 2 patients presented cholestasis. None of them received surgery for the biliary malformation. Eight patients died in the first 20 days of life.

*Choledochal cyst*

Seven (2.8%) patients (3 female, 4 male) [10–12] presented choledochal cyst associated with DA in 5 cases and DS in 2. Four patients (2 DA and 2 DS) had other congenital malformations. The diagnosis of choledocal cyst was received perinatally, during laparotomy for the duodenal malformation repair, by two cases with DA, and at 3 and 7 months, 8 years, and 27 years by the others. The 2 cases with DS were diagnosed of choledocal cyst at 5 and 6 years of age. Four patients presented cholestasis and cholangitis. Five patients underwent surgical correction. One patient died from respiratory distress at 8 months.

*Biliary atresia*

Fourteen (5.6%) patients (3 female, 5 male, and 6 not reported) born with DA, were diagnosed of biliary atresia in the perinatal period in all cases but one premature female, in whom the biliary anomaly was found at 9 months [5,13-22]. Eight children presented other multiple congenital malformations. Seven patients had cholestatic jaundice. Seven patients underwent kasai portoenterostomy and one had a primary transplant. Five patients died in their first year of life.

*Annular pancreas*

Annular pancreas was diagnosed in 146 (58.6%) patients (57 females, 78 males, and 11 not reported), 15 of them with DA, 113 with DS, and 18 with not specified duodenal obstruction [3,23-62]. In 111 patients the presence of annular pancreas was detected in the perinatal period, in 3 at necropsy, in 4 in the first 20 months, and in 28 at a median age of 33 years (range 4–72 years). Other malformations were described in 80 patients. Two newborns and 1 child had pancreatitis. In 18 adults, obstructive symptoms were described, likely more related to the presence of unknown DS. Only 2 adults received division of the annular pancreas in the early years. Twenty-seven patients died, 26 newborns for complications of the surgery and 1 adult.

*Combination of biliary and pancreatic abnormalities*

In the remaining 58 (23.3%) cases (23 females, 31 males, 4 not reported), a combination of biliary and pancreatic abnormalities or a malformation not included in the above groups were diagnosed in 41 DA, 16 DS, and 1 not specified duodenal obstruction [5,10,14,63-66]. The diagnosed was reached at necropsy in 4 patients, perinatally in 31, in the first year of life in 4, at median age of 3 years (range 1–40 years) in 19. Thirty-two of them had other congenital malformations. Thirty presented jaundice, hyperbilirubinemia, or recurrent pancreatitis. Fourteen of them received surgical correction related to the biliary/pancreatic malformations. Fourteen patients died.

## Discussion

The epithelium of the duodenum proliferates in the 4th week of pregnancy, occluding completely the lumen. Four weeks later, the vacuoles start to coalesce forming two parallel channels in the duodenum. At the same time, other two channels appear in the developing biliary system, each of which joins up separately with the duodenal channels [67]. Traditionally, incomplete canalization of the gut has been considered the cause of duodenal stenosis if partial, and of duodenal atresia if complete. When the latter develops between two orifices of the bile ducts, anomalous bifurcated bile duct termination may occur. However, it has been suggested that a “local traffic jam” happening between the duodenum and the entry of the hepaticopancreatic and accessory pancreatic ducts impedes the recanalization of this segment of duodenum, and that terminal branches of the hepatopancreatic ducts could enter the duodenum above and below the atresia [68]. This anomaly of the biliary system may explain the presence on abdominal x-ray of small bowel gas distal to the double bubble of the DA [9]. We found only 24 cases of anomalous bifurcated bile duct detailed in literature [8], mostly associated with DA and all diagnosed perinatally. They did not present symptoms caused by the biliary abnormality, and none of them required surgery for that. One third of the patients died, mostly for complications related to associated congenital malformations. As previously reported [8], it is possible that these abnormalities are far more common, but go undetected at surgery or undescribed in literature. Identification and description of the biliary anatomy is useful for surgeons in order to be aware and to avoid damaging biliary and pancreatic ducts when repairing DA/DS.

The concomitant presence of choldedochal cyst and DA was very rare, and clearly described only in 7 cases in literature. It has been hypothesized that choldedochal cyst is the result of a pancreato-biliary malunion and part of the spectrum of the defective embryogenesis leading to duodenal obstruction [10]. Only in two cases associated with DA the diagnosis was confirmed perinatally; in the remaining patients, it was suggested later, in childhood and in adulthood in one case. It was interesting to note that more than half of the patients had other congenital malformations.

Biliary atresia is an obstructive cholangiopathy of unknown etiology, that was diagnosed in association with congenital duodenal obstruction in 14 cases. It was detected in all cases but one perinatally, and in 8 cases it was associated with more than one other congenital malformations. The high mortality in this group is likely due to the complexity of associated malformations.

Annular pancreas occurs in 1 per 12,000–15,000 live births, causing 8% of duodenal obstruction. It happens in early development of the foregut when the ventral bud fails to rotate behind of the duodenum, leaving pancreatic tissue fully encircling the second portion of the duodenum [51].

It frequently coexists with intrinsic duodenal anomalies (33% of the cases) [69], and, sometimes, it is found accidentally at surgery. It has been suggested that the obstruction of the duodenum in annular pancreas is not the result of intrinsic compression, but is due to a developmental failure [70]. It can become symptomatic in infancy or early childhood, presenting the pattern of a duodenal obstruction. However, some reports had also described presentation of annular pancreas in adults, characterized by abdominal pain, pancreatitis, duodenal ulcer, pancreatic head mass [50,71]. In the vast majority of cases (76%) the diagnosis of annular pancreas was finalized perinatally. In children that subsequently develop recurrent pancreatitis, awareness of the possible association of other biliary and pancreatic malformations [72], as bile duct and pancreatic duct maljunction, should be raised [66]. Pancreatobiliary malunion is characterized by an abnormal connection between the pancreatic and the biliary ducts outside the

duodenum wall and beyond the control of the Oddi's sphincter, leading to regurgitation of bile into the pancreatic duct and consequent pancreatitis [66,72]. On the other hand, when the diagnosis of annular pancreas had been made later, obstructive symptoms related to the associated duodenal obstruction suggested it.

Overall, the most frequent biliary anomaly associated with DA was anomalous bifurcated bile duct, occurring in 21.7% of the cases described in this review. Annular pancreas, on the other hand, accompanied 84.3% of the cases of DS. The presence of other congenital malformations occurred mainly in case of anomalous bifurcated bile duct (58.3% of the cases). The higher percentage (57.1%) of symptomatic patients (cholestasis) was found in cases associated with a choledocal cyst.

Our systematic review presented the current body of information on association between congenital duodenal obstruction and biliary and/or pancreatic malformation. A total of 249 cases were collected from the literature, 84% of which were newborns or children and 16% adults.

The systematic review has higlhlighted some key points. First, the majority (78%) of biliary and pancreatic malformations associated with congenital duodenal obstruction were diagnosed perinatally. Second, biliary and pancreatic malformations were mostly associated to DA, except for annular pancreas which has been described in conjunction with DS in 76% of the cases. Third, the majority (81%) of biliary and pancreatic malformations associated with congenital duodenal obstruction were asymptomatic. When adults were diagnosed with annular pancreas, the symptoms were more related to the unknown duodenal obstruction. Forth, the biliary and pancreatic malformations which are not likely to cause early symptoms, as anomalous biforcated bile duct, were rarely described, but this could be an underestimation of their really incidence. Fifth, other congenital malformations were detected in 55% of the cases. It could be possible that of biliary and pancreatic malformations associated with congenital duodenal obstruction were described in literature mostly in complex cases, and not routinely.

In conclusion, the presence of congenital duodenal obstruction should give warning for a possible association with biliary and/or pancreatic abnormalities. Therefore, careful intraoperative dissection should be performed at DA/DS repair. In addition, persistence of cholestasis, obstructive abdominal symptoms, or recurrent pancreatitis following correction of DA or DS should rise the alert to perform appropriate imaging to look for an association with biliary abnormalities.

## Other information

The review was not registered

A review protocol was not prepared, but all the data extracted are in Table 1 and 2

Financial or non-financial support for the review, and the role of the funders or sponsors in the review: None

## Authors' contribution

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Availability of data: Table 1 and 2

## Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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