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

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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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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:

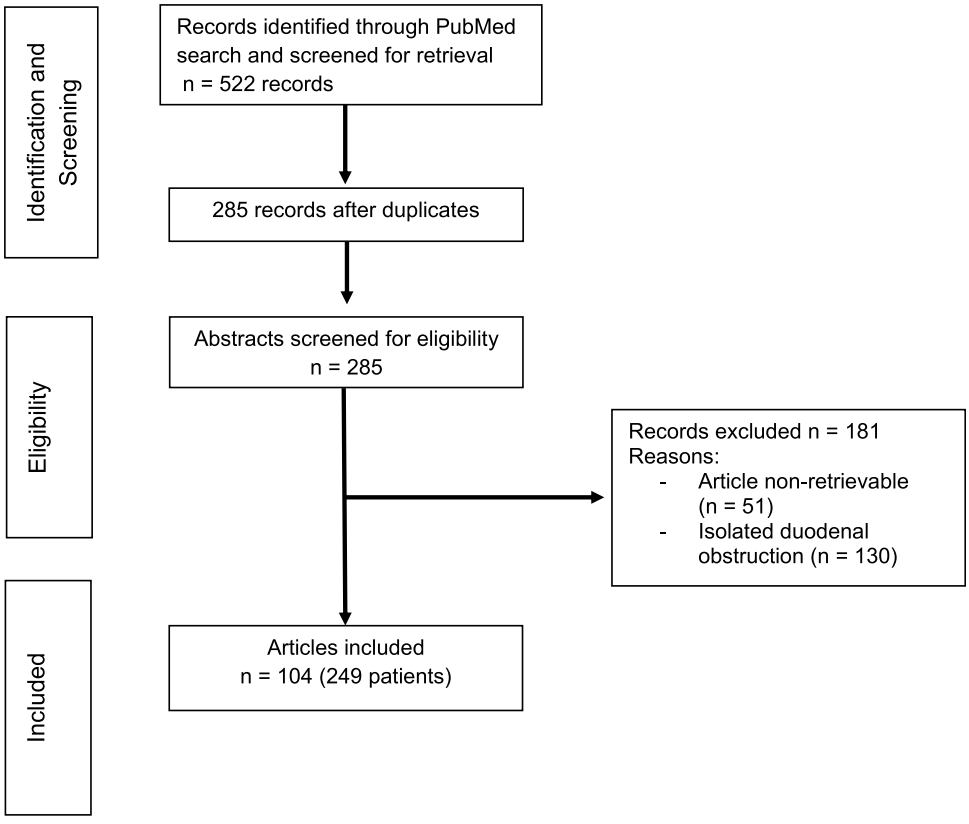

Fig. 1. Study selection flowchart.

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 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 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 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 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 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 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 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 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 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 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 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 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 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 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 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 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 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) 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

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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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