Duodenal Obstruction

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Abstract
The duodenum is the most common site of neonatal intestinal obstruction, accounting for 50% of all intestinal atresias. Duodenal obstruction (DO) is often complicated by prematurity and associated anomalies. Early prenatal ultrasonographic diagnosis of DO allows the mother karyotype analysis for trisomy 21 and other associated anomalies. Cardiac malformations are the major cause of morbidity and mortality in patients with congenital DO.

For most causes of congenital DOs, duodenoduodenostomy via an open approach is the preferred surgical procedure. Vidal from France and Ernst from the Netherlands are credited with the first successful repairs in 1905 and 1914, respectively. Over the last decade, the application of minimally invasive surgical techniques (MIS) and the advent of smaller laparoscopic instruments have expanded the potential of laparoscopy for repair of congenital DO. The first reported laparoscopic repairs of duodenal atresia were by Bax in 2001 and Rothenberg in 2002.

The last retrospective studies comparing the surgical outcome of laparoscopic repair versus open repair found that laparoscopy is a safe and effective technique, and the results including operative time, length of stay, time to full feeding, and complication rate were similar in both groups. The long-term survival rate of patients with DO is excellent and greater than 95% resulting from an early surgical intervention combined with advancement in neonatal intensive care, anesthesia, and nutritional support.

Introduction
Congenital DO is the most common cause of intestinal obstruction in the newborn period, occurring in 1 per 5000–10,000 live births (Best et al. 2012; Choudhry et al. 2009; Haeusler et al. 2002; Rattan et al. 2016). DO is the result of intrinsic lesion, extrinsic lesion, or a combination of both. These pathological lesions can cause complete or incomplete obstruction. Intrinsic DO may be caused by duodenal atresia, stenosis, diaphragm, a perforated diaphragm, or a “wind-sock” web. The “wind-sock” web is a duodenal membrane which is ballooned distally as a result of peristalsis from above (Norton et al. 1992; Rowe et al. 1968). Extrinsic DO may be caused by annular pancreas, malrotation, or preduodenal portal vein (Rattan et al. 2016). Although the annular pancreas forms a constricting ring around the second part of the duodenum (Fig. 1), it is not believed to be the cause of DO (Elliott et al. 1968; Escobar et al. 2004), and there is usually an associated atresia or stenosis in patients with an

Fig. 1 Duodenal obstruction caused by an annular pancreas associated with duodenal stenosis in a postmortem of a 14-week-old fetus with a diagnosis of Down syndrome. ST stomach, DU duodenum, AP annular pancreas (Sweed 2009). (The picture added with the courtesy of Prof. Bronshtein Moshe)
annular pancreas (Girvan and Stephens 1974; Grosfeld et al. 1979; Papandreou et al. 2004). Similarly, preduodenal portal vein has also seldom been reported to be the cause of DO, and it is often associated with other causes of intestinal obstruction such as malrotation or duodenal atresia (Kouwenberg et al. 2008; Singal et al. 2009; Srivastava et al. 2016). Duodenal atresias have been classified into three types: type I bowel continuity but with luminal obstruction or stenosis, type II bowel discontinuity with a connecting bridge of tissue, and type III complete separation with a mesenteric defect (Fig. 2) (Skandalakis and Gray 1994). The reported prevalence of type I is about 92%, type II is 1%, and type III is 7% (Applebaum et al. 2006). Duodenal stenosis is approximately half as prevalent as atresia (Dalla Vecchia et al. 1998).

The obstruction of the duodenum usually occurs distal to the ampulla of Vater. Pre-ampullary obstruction is much less common, occurring in about 20% of cases (Knechtle and Filston 1990). Occasionally there may be a bifid termination of the bile duct with one limb of the duct system opening into the duodenum above the atresia and one below (Komuro et al. 2011; Reid 1973).

Figure 3 shows the wide spectrum of various types of DO. The proximal and distal segments of the duodenum may be separated by a gap (Fig. 3a), be in apposition (Fig. 3b), or be joined by a fibrous cord (Fig. 3c). Other types include duodenal stenosis (Fig. 3d), complete diaphragm (Fig. 3e), a perforated diaphragm (Fig. 3f), a “wind-sock” web (Fig. 3g), and an annular pancreas (Fig. 3h).

**Etiology**

The underlying cause of duodenal atresia remains unknown although its pathophysiology has been well described. Frequent association of duodenal atresia or stenosis with other neonatal malformations suggests that both anomalies are due to a developmental error in the early period of gestation. Tandler (1900) theorized that duodenum is a solid cord during early development, secondary to exuberant epithelial growth, and lumen is formed by vacuoles which coalesce. Further, he suggested that duodenal atresia results from the failure of the solid cord to recanalize. Merrot et al. (2006) were the first investigators to counter the “recanalization theory,” asserting that it failed to explain the different morphological types and other variability seen in patients with duodenal atresia (Teague et al. 2018). Duodenal atresia differs from other atresias of the small and large bowel, which are isolated anomalies caused by mesenteric vascular accidents during later stages of development (Jejuno-Ileal Atresia and Stenosis Essay 367,705 84/84). This theory of vascular disturbance was presented by the classic study of Lauw and Barnard (1955).

No predisposing maternal risk factors are known. Although up to one third of patients with duodenal atresia have Down syndrome (trisomy 21), it is not an independent risk factor for developing duodenal atresia (Applebaum et al. 2006).
In the large California population-based registry of 2.5 million infants, the risk of duodenal atresia was found to be 265 times higher in infants with Down syndrome compared to those without it, and the corresponding frequencies were 46 and 0.12 per 1000 births (Torfs and Christianson 1998). The association between duodenal atresia and Down syndrome suggests an underlying genetic etiology. In mice, interruption of fibroblast growth factor 10 (FGF10) gene signaling results in duodenal atresia in 30–50% of embryos supporting genetic etiology (Teague et al. 2018).

Although DO is usually not regarded as a familial condition, there have been several reports of familial cases (Gahukamble et al. 1994; Gross et al. 1996; Markljung et al. 2012; Okti et al. 2005) and a very rare group of hereditary multiple intestinal atresias with fatal outcome (Lambrecht and Kluth 1998). Markljung et al. reported recently on a new familial case of annular pancreas and found one microduplication on chromosome 6q24 by array-based comparative genomic hybridization (CGH) shared by the affected mother and son (Markljung et al. 2012). This microduplication may be a causative aberration or present a risk factor for the development of annular pancreas and duodenal atresia.

### Associated Malformations

There is a high incidence (approximately 50%) of associated anomalies in patients with intrinsic DO, especially Down syndrome which occurs in about 30% of these patients (Sweed 2011; Puri 1981; Young and Wilkinson 1968).

Table 1 presents the overall prevalence and distribution of associated anomalies of duodenal atresia. The data are the collected statistics of 1759 patients with DO from a dozen large series.
The associated anomalies in order of frequency are Down syndrome (28%), annular pancreas (23%), congenital heart disease (22.6%), malrotation (20%), esophageal atresia (8.5%), genitourinary malformations (8%), anorectal anomalies (4.4%), and other bowel atresias (3.5%).

Vertebral anomalies were reported to range between 2% (Bailey et al. 1993) and 37% (Atwell and Klidkjian 1982) in these patients. Reports of duodenal atresia have also shown a low incidence of musculoskeletal anomalies (Pulkkinen et al. 1997).

Other rare anomalies include Cornelia de Lange syndrome (Bailey et al. 1993), chromosomal abnormalities (Sweed 2011), multiple intestinal abnormalities (Morikawa et al. 2009), choledochal cyst (Iwai et al. 2009; Sugimoto et al. 2004), immunodeficiency (Moore et al. 1996), tracheomalacia (Kimble et al. 1997), and situs inversus (Nawaz et al. 2005).

The complex cardiac anomalies among all other associated malformations are the major cause of morbidity and mortality in patients with duodenal atresia (Choudhry et al. 2009; Dalla Vecchia et al. 1998; Escobar et al. 2004; Piper et al. 2008). Dalla Vecchia et al. attributed all the operative mortality (4%) to associated complex congenital heart anomalies in a group of 138 patients with DO in a 25-year survey. Two other important factors affecting higher morbidity and mortality of these patients are prematurity and low birth weight (Escobar et al. 2004; Gourevitch 1971; Hancock and Wiseman 1989; Piper et al. 2008). The mortality rate is even higher in neonates born with three or more anomalies of the VACTERL association with an overall survival rate of 40–77% (Iuchtman et al. 1992; Muraji and Mahour 1984; Weber et al. 1980). Spitz and colleagues reported the combination of esophageal and duodenal atresias as particularly lethal, with mortality rates ranging from 67% to 94% in various series (Spitz et al. 1981). Jackson et al. inferred that the majority of these deaths are caused by failure to recognize the second abnormality preoperatively (Jackson et al. 1983).

Maternal polyhydramnios has been reported to be present in 17–75% of cases of duodenal atresia (Cohen-Overbeek et al. 2008; Hancock and Wiseman 1989; Murshed et al. 1999) and is the most common ultrasonographic finding in fetuses with intrinsic DO (Irving 1990). Ultrasound is usually performed for suspected fetal or maternal abnormalities when polyhydramnios or a large-for-date pregnancy is established. Although the majority of cases are diagnosed during the seventh or eighth month of gestation (Bronshtein et al. 2008), sonographic detection of duodenal atresia was reported as early as 12 gestational weeks by Tsukerman et al. (1993) and 19 weeks by Romero (1988).

There has been an increase in prenatal ultrasonographic diagnosis of duodenal atresia during the last three decades, from 14% between the years 1972 and 1991 (Grosfeld and Rescorla 1993) to 18% (Akhtar and Guiney 1992) to the high rate of 57% for the period of 1991–1995 (Murshed et al. 1999).

The prenatal sonographic diagnosis relies on the demonstration of the “double bubble” sign, which is due to simultaneous distension of the stomach and the first part of the duodenum (Fig. 4). In many cases this sonographic sign is observed in the second half of pregnancy probably due to hydrostatic pressure needed to dilate the duodenum and also to the degree of the DO.

Visualization of a fluid-filled double bubble (Fig. 4) on prenatal ultrasound scan is associated with DO secondary to intrinsic or extrinsic lesion. This sonographic finding is known to have a low

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**Table 1** The incidence of associated congenital anomalies (%) (Collected statistics) (*N* = 1759 patients). (Data from Sweed 2009)

| Associated anomaly                                      | %    |
|--------------------------------------------------------|------|
| Down syndrome                                          | 28.2 |
| Annular pancreas                                       | 23.1 |
| Congenital heart disease                               | 22.6 |
| Malrotation                                            | 19.7 |
| Esophageal atresia and tracheoesophageal fistula        | 8.5  |
| Genitourinary                                          | 8.0  |
| Anorectal                                              | 4.4  |
| Other bowel atresia                                    | 3.5  |
| Others                                                 | 10.9 |

(Sweed 2011). The associated anomalies in order of frequency are Down syndrome (28%), annular pancreas (23%), congenital heart disease (22.6%), malrotation (20%), esophageal atresia (8.5%), genitourinary malformations (8%), anorectal anomalies (4.4%), and other bowel atresias (3.5%).
false-positive rate. Zimmer and Bronstein have reported that in a few cases, it may represent a transient finding in an otherwise healthy fetus (Zimmer and Bronstein 1996). It is possible that intestinal peristalsis in a fetus may show transient dilatation suggesting DO (Bronshtein et al. 2008). On the ultrasound examination, it is also important to demonstrate the continuity between the gastric and duodenal bubbles (Fig. 4) to exclude other causes such as choledochal cyst which lacks such communication (Casaccia et al. 2002; Lawrence et al. 2000) or duodenal duplication (Malone et al. 1997).

Often other anomalies can also be diagnosed by ultrasound. Kawana et al. (1989) and Pameijer et al. (2000) reported on the ultrasonic prenatal diagnosis of a fetus with combined duodenal and esophageal atresias associated with VACTERL anomalies. Prenatal ultrasonographic diagnosis of annular pancreas has also been reported showing the coincidence of the double bubble sign together with hyperechogenic bands around the duodenum (corresponding with the tissue of annular pancreas) (Dankovcik et al. 2008).

Hancock and Wiseman investigated the impact of antenatal diagnosis of congenital DO in a series of 34 infants, 15 of whom were diagnosed by antenatal ultrasound (Hancock and Wiseman 1989). They concluded that although surgery was performed sooner, the outcome of infants with DO was not changed by providing an antenatal diagnosis. However, the antenatal diagnosis of DO influenced parents positively in coping with the anomaly, because it allowed them time to prepare for the medical and surgical interventions required after the birth of their infant. These authors also emphasize that a normal ultrasound in the presence of polyhydramnios does not rule out the diagnosis of DO and is an indication for repeated sonography. Cohen-Overbeek et al. have also reported on 91 cases diagnosed with isolated or non-isolated DO. They found that the outcome of prenatally and postnatally diagnosed DO is not essentially different despite the fact that more prematurity and a lower birth weight were observed in the former (Cohen-Overbeek et al. 2008).

The rapid advancement in imaging technology, including magnetic resonance imaging (MRI), should allow for diagnosis during the first and early second trimester, enabling abortion (Tsukerman et al. 1993). Alternatively, early prenatal diagnosis of DO should lead to karyotype analysis for prenatal screening for trisomy 21 and other associated anomalies (Grosfeld and Rescorla 1993; Keckler et al. 2008; Singh et al. 2004).

The prenatal diagnosis allows the mother the opportunity to receive counseling and to consider delivery at or near a tertiary care facility that is able to care for infants with gastrointestinal anomalies (Haeusler et al. 2002).

Clinical Presentation and Diagnosis

The presenting symptoms and signs are the result of high intestinal obstruction. About half of these patients are premature and low birth weight infants (Dalla Vecchia et al. 1998; Escobar et al. 2004; Piper et al. 2008). Vomiting is the most common symptom and is usually present on the first day of life. Since 80% of the obstructions are located in the post-ampullary region of the
duodenum, vomitus in the majority of cases is bile-stained. In supra-ampullary atresia it is non-bilious. Orogastric aspiration also yields significant volumes of bile-stained gastric fluid. There is minimal or no abdominal distension because of the high level of obstruction. The infant may pass some meconium in the first 24 h of life and thereafter constipation may develop. Dehydration with weight loss and electrolyte imbalance (hypokalemic/hypochloremic metabolic alkalosis) soon follows if the diagnosis is done late and if fluid and electrolyte losses have not been adequately replaced (Kilbride et al. 2010). Incomplete DO usually leads to the delayed onset of symptoms. Infants with duodenal stenosis and partial bowel obstruction may escape detection of an abnormality soon after birth and may proceed into childhood or rarely into adulthood before a partial obstruction is noted (Escobar et al. 2004; Grosfeld and Rescorla 1993).

The diagnosis of DO is confirmed on X-ray examination. An abdominal radiograph will show a dilated stomach and duodenum, giving the characteristic appearance of a double bubble sign (the stomach and the proximal duodenum are air filled) with no gas beyond the duodenum (Fig. 5a–c). In partial DO, a plain film of the abdomen will show a double bubble appearance, but there is usually some air in the distal intestine (Fig. 6). Occasionally in cases of duodenal atresia, air may be seen distal to the site of obstruction due to associated bile duct bifurcation (Knechtle and Filston 1990). Radiographic findings in patients with annular pancreas are usually indistinguishable from duodenal atresia or stenosis.

In some cases of partial DO, plain films may be normal. Upper gastrointestinal tract contrast radiography is indicated in these patients to establish the cause of incomplete DO. This may show a stenotic segment of duodenum with dilatation of the proximal segment or a sharp termination of the dilated segment, indicating a perforated diaphragm (Fig. 7).

Incomplete DO usually leads to delayed onset of symptoms, and the diagnosis of duodenal diaphragm with a central aperture is sometimes delayed for months or even years (Melek and Edirne 2008; van Rijn et al. 2006; Vaos and Misiakos 2010). Mikaelsson et al. reported on the late diagnosis and treatment of 8 out of 16 patients with membranous duodenal stenosis. Their patients were diagnosed and operated at 1 month to 4 years of age (Mikaelsson et al. 1997). Occasionally a duodenal diaphragm may be stretched and ballooned distally, giving the “wind-sock” appearance on a contrast study (Fig. 8; Eustace et al. 1993).

The most important differential diagnosis of DO is DO caused by malrotation resulting in extrinsic compression related to Ladd’s bands across the duodenum, or volvulus of the midgut loop, although this is rare. Midgut volvulus may result in gangrene of the entire midgut within hours, and thus diagnostic investigation is urgently required, though the symptoms may relent because the obstruction may be incomplete or intermittent in malrotation. Part of these extrinsic obstructions exhibits the double bubble sign with distal air on plain film, while the majority can be identified from the coil spring appearance of small bowel volvulus following barium injection (Eustace et al. 1993). However, Samuel et al. observed that volvulus neonatorum was not encountered in neonates with duodenal atresia and stenosis who had associated malrotation. They suggested that DO could perhaps be a floodgate that prevents volvulus in these children (Samuel et al. 1997).

Preduodenal portal vein is a rare anomaly and generally asymptomatic. It is a rare cause of DO and often coexists with other anomalies resulting in bowel obstruction (Katari et al. 1998; Mordehai et al. 2002; Singal et al. 2009). In most of these patients, it is impossible to diagnose preduodenal portal vein prior to surgery.

The wide variety of additional congenital anomalies with special emphasis on cardiac malformation, often severe (Keckler et al. 2008; Piper et al. 2008), make preoperative diagnosis imperative. Anterior-posterior and lateral chest and abdominal radiographs ascertaining visualization of the entire spine should also be performed.

Soon after the X-ray, cardiac and renal ultrasound should be carried out routinely in all these babies. A micturating cystourethrogram should be
performed in those babies with abnormal urogenital ultrasound or an associated anorectal anomaly. Rectal biopsy should be taken in babies with constipation and the combination of Down syndrome and duodenal atresia, to exclude Hirschsprung’s disease (Kimble et al. 1997).

Fig. 5 (a) Abdominal radiograph showing grossly distended stomach and duodenum with “double bubble” sign with no air beyond the duodenum. GB gastric bubble, DB duodenal bubble (Sweed 2011). (b) Abdominal radiograph showing the “double bubble” sign. In this case duodenal bulb is more prominent than the gastric bulb. At operation duodenal membrane was found and excised. GB gastric bubble, DB duodenal bubble (Sweed 2011). (c) Duodenal atresia evident on upper gastrointestinal radiograph contrast study. S stomach, D duodenum (Sweed 2011)

Preoperative Management

Although duodenal atresia is a relative emergency, the patient should not be rushed to the operating room until the infant’s hemodynamic and fluid and electrolyte status is stable. If the clinical history and findings on physical
examination indicate that the baby is in no distress
and the radiographs are consistent with the usual
presentation of duodenal atresia with no air beyond
the second bubble (excluding malrotation), opera-
tion should be performed on an elective basis.

An orogastric tube decompresses the stomach,
and intravenous fluid resuscitation can be initi-
ated. Blood samples for electrolyte determination
should be obtained, and any derangements should
be corrected. Prolonged vomiting can result in a
hypokalemic hypochloremic metabolic alkalosis.
Passage of the orogastric tube rules out esopha-
geal atresia, and careful inspection of anal defect
variants of imperforated anus should be obtained.

Care is taken to preserve body heat and avoid
hypoglycemia, since many of these newborn
patients are premature and small for date
(Murshed et al. 1999). Very low birth weight
infants or those with respiratory distress syndrome
and associated severe anomalies, e.g., congenital
heart disease, may need occasionally special pre-
paration such as resuscitation and ventilation.

Operation

Duodenoduodenostomy is the procedure of
choice for patients with duodenal atresia, stenosis,
and annular pancreas (Dalla Vecchia et al. 1998;
Weber et al. 1986; Wesley and Mahour 1977).

Duodenoduodenostomy can be performed in
either “diamond-shaped” (proximal transverse to
distal longitudinal) anastomosis as described by
Kimura (Fig. 9a, b; Kimura et al. 1977) or side-to-
side fashion (Fig. 10). The “diamond-shaped”
duodenoduodenostomy has been reported to
allow earlier feeding, earlier discharge, and good
long-term results (Kimura et al. 1990; Upadhyay
et al. 1996).

Bax et al. (2001) and Rothenberg (2002)
reported on the first case and the first series,
respectively, on the of laparoscopic management
of DO. They indicated that laparoscopic approach
has proven to be safe and effective and represents an alternative to the open procedure. They also emphasized that this minimal invasive surgical technique should be used only if the surgeon has appropriate instruments and suturing and laparoscopic skills (Bax et al. 2001; Kay et al. 2009; Rothenberg 2002).

**Incision**

The baby is placed supine on the table with a small roll under his upper abdomen on a warming blanket. Endotracheal anesthesia is used. The abdominal skin is prepared by cleaning with prewarmed povidone-iodine.

A transverse supra-umbilical abdominal incision is made 2 cm above the umbilicus starting in the midline and extending laterally into the right upper quadrant for about 5 cm. The abdominal muscles are divided transversely with cutting diathermy, and the peritoneal cavity is opened in the line of incision.

**Exploration and Identification of Pathology**

After exposing the peritoneal cavity, the surgeon inspects the entire bowel for the presence of other bowel anomalies. There may be an associated annular pancreas, malrotation (in about one third of the patients), or in rare cases, preduodenal portal vein. If the colon is in normal position, malrotation is probably not a coexisting factor.

The stomach and first portion of the duodenum are usually thickened and dilated. The liver is carefully retracted superiorly. The ascending colon and the hepatic flexure of the colon are

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**Fig. 8** “Wind-sock” web. Dilated duodenum demonstrated with duodenal membrane ballooned distally, giving characteristic “wind-sock” appearance. Reflux of contrast medium into pancreatic and common bile duct is seen. (As appeared in Fig. 49.10, page 474 of the textbook “Newborn Surgery” 3rd ed. by Puri P, Sweed 2009)

**Fig. 9** Diamond-shaped duodenoduodenostomy. (a) A transverse incision is made in the distal end of the proximal dilated duodenum, and a longitudinal incision is made in the smaller limb of the duodenum distal to the occlusion. (b) A single-layer anastomosis using interrupted 5/0 Vicryl sutures with posterior knots tied inside the posterior wall of the anastomosis and anterior knots tied outside the anterior wall is performed.
mobilized medially and downwards to expose the dilated duodenum (Sweed2006).

The duodenum is then adequately mobilized and freed from its retroperitoneal attachments – Kocher maneuver. Great care must be exercised not to dissect or manipulate either segment of the duodenum medially, to avoid injury to the ampulla of Vater or the common bile duct. The tube in the stomach is then passed distally into the dilated duodenum and helps to locate the point of obstruction and determine if a “wind-sock” deformity is present (Fig. 3g).

The type of atresia as well as any pancreatic abnormality (e.g., annular pancreas) is noted. In patients with an annular pancreas, the pancreatic tissue should never be divided and should always be bypassed. The duodenum distal to the site of obstruction is small and decompressed. The requirements for distal mobilization vary according to the location of the atresia and to the gap between the two segments (Fig. 2). If necessary, the ligament of Treitz is divided, and mobilization and displacement of the distal duodenum are performed behind the superior mesenteric vessels, thus allowing a satisfactory anastomosis to be performed without any tension.

**Fig. 10** Side-to-side duodenoduodenostomy. (a) An upper transverse abdominal incision. (b) Parallel incisions of about 1 cm are made in the proximal and distal duodenum. (c) The anastomosis is performed using single-layer interrupted 5/0 Vicryl sutures. (As appeared in Fig. 49.10, page 474 of the textbook “Newborn Surgery” 3rd ed. by Puri P, Sweed 2009)

**“Diamond-Shaped” Duodenoduodenostomy**

After abdominal exploration, the duodenum is adequately mobilized. With two traction sutures, the redundant wall of the proximal duodenum is pulled downward to overlie the proximal portion of the distal duodenal segment. A transverse incision is made in the distal end of the proximal duodenum, and a longitudinal incision is made in the smaller limb of the duodenum distal to the occlusion.

These are made in such a position as to allow good approximation of the openings without tension.

The papilla of Vater is located by observing bile flow. This is performed by gentle compression of the gallbladder.

The orientation of the sutures in the “diamond-shaped” anastomosis and the overlapping between the proximal transverse incision and the distal longitudinal incision are shown in Fig. 9a, b.

Additionally, an 8 French Foley catheter should be passed proximally into the stomach and distally into the jejunum and pulled back with the balloon inflated, to ensure that no additional web or a “wind-sock” deformity is
overlooked. The distal duodenum can be distended to a larger size during this maneuver facilitating the anastomosis. Before pulling back the catheter from the distal duodenum, the surgeon should inject 30–40 ml of warm saline through the catheter to rule out distal atresias of the distal small bowel. The catheter is then removed.

A single-layer anastomosis using 5/0 or 6/0 Vicryl sutures with posterior knots tied inside the posterior wall of the anastomosis and interrupted sutures with anterior knots tied outside the anterior wall. Before completion of the anterior part of the anastomosis, a 5F silicon nasojejunal transanastomotic feeding tube maybe passed down into the upper jejunum for an early postoperative enteral feeding (Hall et al. 2011) using the same insertion technique as was reported for patients who underwent surgical repair for esophageal atresia and tracheoesophageal fistula (Sweed et al. 1992). Others, however, do not use the nasojejunal tube because they suggest that it may delay the commencement of oral feeding (Kimura et al. 1990). Hall et al. reported recently that a transanastomotic tube significantly shortens time to full enteral feeds in infants with congenital DO as well as significantly reducing the need for central venous access and parenteral nutrition (Hall et al. 2011). Then the right colon is returned to its former position so that the mesocolon covers the anastomosis. The Ladd procedure with “inversion appendectomy” is performed in patients with malrotation (Grosfeld and Rescorla 1993). In these patients, the cecum should be placed in the left lower quadrant to reduce the risk of midgut volvulus.

The wound is closed in layers: the peritoneum and posterior fascia and the anterior fascia by two layers using continuous 4-0 Vicryl. The skin is closed with running intracuticular suture using 5-0 Vicryl.

### Side-to-Side Duodenoduodenostomy

The dilated proximal duodenum and the distal collapsed duodenum are approximated using two stay sutures (5-0 Vicryl). Then parallel incisions with a length of about 1 cm are made in the proximal and distal duodenum (Fig. 10). An 8 French Foley catheter should be inserted both to the proximal dilated duodenum and to the distal collapsed duodenum in order to rule out “wind-sock” membrane and distal atresias similarly as described in the “diamond-shaped” duodenoduodenostomy.

The posterior layer of anastomosis is completed using interrupted 5/0 Vicryl sutures.

At this stage, a transanastomotic 5 Fr gauge silastic nasojejunal tube may be inserted for an early enteral feeding.

The anastomosis is then completed using interrupted 5-0 Vicryl sutures for the anterior layer. The abdomen is closed in the same manner as described in the “diamond-shaped” duodenoduodenostomy.

In premature infants, some surgeons prefer to perform a gastrostomy and insert the transanastomotic silicon tube via the gastrostomy. The tip of the tube should be well down in the jejunum as to decrease the chance of it becoming displaced.

### Operative Technique for Duodenal Web

A longitudinal incision is performed above the “transitional zone” between the wide and narrow segments of the duodenum (Fig. 11a), and the duodenum is opened. The membrane usually is located in the second part and occasionally in the third portion of the duodenum. It can be complete or have a hole. Anatomically, the ampulla of Vater may open directly into the medial part of the membrane, or posteriorly close to it; thus the close relationship of the membrane to papilla of Vater makes its identification mandatory, before excision of the web. Excision of the web should proceed from the lateral duodenal wall, leaving the medial third of the wall intact to avoid damaging the sphincter of Oddi or the ampulla of Vater and continue leaving a circumferential rim of tissue of 1–2 mm (Fig. 11b, d). The resection line is then oversewn using continuous sutures of Vicryl 5/0, and the duodenum is closed.
transversely in one layer using Vicryl 5/0 (Fig. 11c). Because of the pitfalls in cases of the lax membrane that may bulge downward distally into the distended duodenum (the so-called windsock phenomenon), and in order to avoid missing the anomaly, before closure of the duodenum, the distal patency of the distal duodenum must be verified by inserting an 8 French Foley catheter through duodenotomy (Fig. 11e, f).

The experience with fiber optic duodenoscopy indicates the usefulness of the technique for both the diagnosis and nonoperative management of duodenal membrane (Beeks et al. 2009; Bittencourt et al. 2012; Okamatsu et al. 1989). However, based on reports describing anomalous entry of the pancreatobiliary channels (Adams 1986), the delineation of the ducts at endoscopic retrograde cholangiopancreatography (ERCP) may be necessary prior to endoscopic intervention.

Recently, Bittencourt et al. reported on three female patients aged between 9 and 12 months born with duodenal membrane who were treated...
successfully by two endoscopic sessions. The first and second sessions of endoscopic treatment included dilatation and resection of the membrane, respectively, and were carried out without complications (Bittencourt et al. 2012).

Most surgeons however believe that a duodenotomy is preferable to the potential risk of inadvertent pancreatic or bile duct injury (Adams 1986).

**Laparoscopic Management of DO**

The application of minimally invasive surgical techniques (MIS) for the correction of congenital anomalies has increased significantly over the last 10 years. The ability to perform delicate dissection and intracorporeal anastomosis has broadened the scope of entities that can be approached including neonatal DO. Although most neonatal conditions presenting with bowel obstruction present a difficult problem for laparoscopy because of the dilated bowel and limited abdominal cavity, this is not the case in duodenal atresia. The entire small and large bowel is decompressed, and there is an excellent exposure of the proximal duodenum (Bax et al. 2001; Kay et al. 2009).

For the laparoscopic approach (Fig. 12), neonatal laparoscopic instruments (3 mm) and trocars are used. The patient is positioned supine at the end of the operating table. The operating surgeon stands at the patient’s feet. The abdomen is insufflated through a 5-mm umbilical port, for a 30° laparoscope. 3-mm and 5-mm ports are placed in the right lower quadrant and left upper quadrant, respectively. The left upper quadrant port is placed for the introduction of suture.

The duodenum is then kocherized, the type of obstruction becomes easily visible, and the dilated proximal and decompressed distal segments are identified (Rothenberg 2002). A proximal transverse and distal longitudinal duodenotomy is then made. As with the open repair, stay sutures are placed at each corner to facilitate the anastomosis. A diamond-shaped anastomosis is performed with either a separate running suture for the posterior and then anterior wall or single interrupted stitches of Vicryl. Intracorporeal knot tying is used. An extra port can be placed in the right upper quadrant to help retract the liver and set up the anastomosis. Alternatively, the apical stitch can be tied and brought out through the abdominal wall to assist with retraction and align the enterotomies for the anastomosis. The distal bowel is examined in all cases to ensure that there are no obvious secondary atresias. Once the anastomosis is completed, the ports are removed, and the sites are closed with absorbable sutures.

The main benefits of laparoscopic approach for the treatment of duodenal atresia are the excellent visualization of the obstruction and the ease of the anastomosis. However, the possible disadvantage of this approach may be that evaluation of the distal bowel for other artretic segments is more difficult to accomplish and, if not specifically evaluated, it is feasible that a malrotation can be missed (Hill et al. 2011). The bowel can be inspected visually for distal obstructed segments, but internal webs may be more difficult to see.

Hill et al. (2011) reported recently on their results comparing 22 patients with DO treated by laparoscopy and 36 patients treated by traditional laparotomy during a 9-year period (2001–2010). They found no difference between groups in time to full feeding, postoperative length of stay, and complication rate. They found that the operative time was slightly longer in the laparoscopic group (median time 116 min vs. 103 min); however, laparoscopic management appeared to allow a
shorter postoperative ventilator requirement. Six patients (26%) of the laparoscopic group were converted to open exploration because of unclear anatomy.

The experience with laparoscopic duodenoduodenostomy (Oh et al. 2017; Spilde et al. 2008; Valusek et al. 2007) demonstrates that it can be performed safely and successfully in the neonate with excellent short-term outcomes. Surgeons with experience in advanced laparoscopic techniques can learn the laparoscopic duodenoduodenostomy and have good results.

**Postoperative Care**

The baby is returned to an incubator (or radiant heat cot) at the thermoneutral temperature for its size and maturity. An intravenous infusion of the dextrose/saline is continued in the postoperative period, and further fluid and electrolyte management depends on clinical progress, loss by gastro-duodenal aspiration, and serum electrolyte levels. Postoperatively, patients may have a prolonged period of bile-stained aspirate through the nasogastric tube, which is mainly due to the inability of the markedly dilated duodenum to produce effective peristalsis. Enteral feeding through the trans-anastomotic feeding tube is generally started within 24–48 h postoperatively.

The commencement of oral feeding depends on the decrease of the gastric aspirate and may be delayed for several days and occasionally for 1–2 weeks or longer. Once the volume of the gastric aspirate decreases, the feeding tube is withdrawn, and the infant can be started on oral feeding.

Spigland and Yazbeck (1990), in their follow-up of 33 neonates, found that bowel transit was established for an average of 13.1 days, 7.5 days after partial web excision, 12.4 days following duodenoduodenostomy, and 15 days after duodenojejunostomy. Spilde et al. (2008) recently reported that the time to initial feeding was 11.3 days for patients with an open repair of DO compared to 5.4 days for those who were treated by laparoscopic approach. They also found that the average time to full oral intake was 16.9 days for the open group compared to 9 days for the laparoscopic group.

**Management of Persistent Megaduodenum by Duodenoplasty**

The deformity and dysfunction of the first part of the duodenum – the megaduodenum – are the causes of well-known morbidity (Ein and Shandling 1986; Spigland and Yazbeck 1990), and occasionally these patients require duodenoplasty (Dewan and Guiney 1990). The malfunction of the greatly dilated gut and the absence of effective peristalsis were demonstrated by Nixon in the small bowel (Nixon 1960), but the same phenomenon is thought to occur in the dilated duodenum proximal to the duodenal atresia. Several techniques of duodenoplasty have been described, and in all, it is of the utmost importance to visualize and identify the ampulla of Vater within the duodenal lumen prior to resection and tapering of the duodenum. Hutton and Thomas have reported success by extensive tapering duodenoplasty (Hutton and Thomas 1988). Adzick et al. (1986) and Grosfeld and Rescorla (1993) emphasized the merit of tapering duodenoplasty at the primary operation of neonates with dilated duodenum, to improve the immediate postoperative gastrointestinal function and the prevention of further development of megaduodenum. Other techniques include resection and suturing (Weisgerber and Bourreau 1982), resection and stapling (Adzick et al. 1986), and elliptical seromuscular resection (Kimura et al. 1996).

However, refashioning the anastomosis or bypass techniques usually fail (Ein et al. 2000; Young et al. 1993). Another technique of subtotal duodenal resection with reconstruction of the duodenum by the proximal jejunum as an onlay patch was demonstrated in two children. In this technique, the diseased duodenal wall is completely removed, except for the area of the ampulla of Vater, and the duodenum is reconstructed by the jejunum (Endo et al. 1998).
Outcome and Long-Term Results

The survival of babies with DO has gradually improved over the last 40 years (Table 2; Bailey et al. 1993; Cohen-Overbeek et al. 2008; Dalla Vecchia et al. 1998; Hill et al. 2011; Kilbride et al. 2010; Wesley and Mahour 1977). All agree that the three main factors contributing to the mortality rate in this group of patients are high incidence of associated anomalies, especially severe cardiac malformations, prematurity, and low birth weight (Choudhry et al. 2009; Escobar et al. 2004; Grosfeld and Rescorla 1993). In a recent review covering 45 years (1951–1995) of management of DO, Murshed et al. (1999) found that in the first 15 years, survival reached 51%, in the next 15 years it was 80%, and in the last 15 years 95%.

During the latest period, mortality was almost entirely the consequence of associated anomalies.

Dalla Vecchia et al. reported a relatively low rate of postoperative complications in a series of 138 infants (Dalla Vecchia et al. 1998). The early complication rate included anastomotic obstruction in 3%, congestive heart failure in 9%, prolonged adynamic ileus in 4%, pneumonia in 5%, and wound infection in 3%.

Late complications included adhesive bowel obstruction in 9%, megaduodenum and duodenal dysmotility that required tapering duodenoplasty in 4%, and gastroesophageal reflux requiring surgery in 5%.

Weber et al. (1986) reported the complication rate and morbidity of 3 methods of technical repair in a group of 41 newborns with duodenal atresia. The three techniques were (1) side-to-side duodenoduodenostomy, (2) side-to-side duodenojejunostomy (which is rarely performed today), and (3) diamond-shaped duodenoduodenostomy. There was no difference in the complication rate, but the “diamond-shaped” technique was found to be superior for repair, resulting in earlier feeding and discharge. Kimura et al. reported on their experience with 44 patients with the diamond-shaped technique (Kimura et al. 1990), without the use of gastrostomy or trans-anastomotic tube, and found a very low rate of complications and good long-term results.

Long-term results of congenital DO were reported by Kokkonen et al. (1998), who studied 41 patients aged 15–35 years. They found that growth and development, including body weight, were satisfactory. Although the great majority was symptom-free, on barium meal examination, all but two had abnormal findings, including mega duodenum in nine cases. They concluded that some gastrointestinal disturbances are common, even in asymptomatic patients, and careful follow-up is important. Salonen and Makinen reported previously on their experience in a small group of nine patients at age 3–21 years (Salonen and Makinen 1976) and found, in contrast, a normal barium meal in all patients except one. Their result was similar to the documentation by Kimura et al. (1990) with the diamond-shaped technique.

Ein et al. encountered five patients with late complications of duodenal atresia repair that appeared suddenly between the ages of 6 months and 24 years. The duodenal repair was functionally obstructed – caused by proximal, dilated

Table 2

| Author/year                  | No. of patients | % survival |
|-----------------------------|-----------------|------------|
| Girvan and Stephen (1974)   | 158             | 67         |
| Wesley and Mahour (1977)    | 72              | 74         |
| Hancock and Wiseman (1989)  | 34              | 94         |
| Akhtar and Guiney (1992)    | 49              | 94         |
| Bailey et al. (1993)        | 138             | 93         |
| Grosfeld and Rescorla (1993)| 103             | 95         |
| Dalla Vecchia et al. (1998)| 138             | 86         |
| Cohen-Overbeek et al. (2008)| 91             | 91         |
| Choudhry et al. (2009)      | 65              | 96         |
| Kilbride et al. (2010)      | 51              | 98         |
| Kay et al. (2009)           | 17              | 100        |
| Hill et al. (2011)b         | 58              | 100        |

aLaparoscopic duodenoduodenostomy (Years: 2004–2008), \(N = 17\) pts
bLaparoscopic versus open repair of duodenal obstruction, (Years: 2001–2010), Lap. Group, \(N = 22\) pts.; open group, \(N = 36\) pts. One patient in the Lap group who had Down syndrome and duodenal and jejunal atresias died because of sepsis 5 months after the initial operation.
duodenal atony. Plication of the dilated atonic proximal duodenum was curative (Ein et al. 2000).

Recently, Son and Kein (2017) compared the results of laparoscopic versus open surgery in the management of duodenal obstruction in 112 neonates. They reported that the laparoscopic treatment is associated with lower postoperative morbidity, shorter recovering time and postoperative hospital stay, and better postoperative cosmesis than open surgery. Mentessidou and Saxena (2017) and Chung et al. (2017) performed systematic reviews and found that laparoscopic repair was of comparable safety and efficacy as the open repair for duodenal atresia.

**Conclusion and Future Directions**

Duodenal obstruction is a common intestinal atresia, often complicated by prematurity and associated anomalies. Currently, duodenoduodenostomy is the treatment of choice and can be performed via open approach or minimally invasive. The last retrospective studies comparing the surgical outcome of laparoscopic repair versus open repair found that laparoscopy is a safe and effective technique, and the results including operative time, length of stay, time to full feeding, and complication rate were similar in both groups (Chung et al. 2017). However, for those preferring laparoscopic DO repair, duodenojejunostomy might be a feasible alternative as it is easier to perform and has equal clinical outcomes compared to duodenoduodenostomy (Zani et al. 2017).

Over the last few decades, advancements in neonatal intensive care, parenteral nutrition, management of associated anomalies, and improvements in operative technique including video equipment, smaller instruments, and better postoperative care have improved the outlook for patients born with duodenal atresia and stenosis. Mortality today has been reduced to 5–10% and is now related mostly to associated cardiac anomalies.

**Cross-References**

- Esophageal Atresia
- Jejuno-Ileal Atresia and Stenosis
- Malrotation

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