Chapter

Duodenal Atresia

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Abstract

Congenital duodenal obstruction (CDO) is a common surgical disease in newborns accounting for up to 50% of all intestinal atresia cases. Duodenal atresia is a form of CDO that occurs due to failure of the solid core of the duodenum to recanalize during fetal development. Patients with duodenal atresia require complex surgical reconstruction. In 1997, Kimura et al. reported a technique of a diamond-shaped anastomosis revolutionizing the management of duodenal atresia. Initially, this repair required a laparotomy. Increasingly, laparoscopy has been used as an alternative approach for duodenal atresia repair. Laparoscopic reconstruction of duodenal atresia is a challenging procedure requiring creation of a delicate anastomosis in a small workspace.

Keywords: duodenal atresia, duodenal obstruction, laparoscopy, duodenal anastomosis, minimally invasive, duodenal web, duodenal membrane, transoral

1. Introduction

Congenital duodenal obstruction (CDO) is a common surgical anomaly in newborns [1, 2] that can be diagnosed prenatally and requires careful planning for surgical repair after birth. This chapter focuses on the surgical management duodenal atresia, a common form of CDO. With the rising popularity of laparoscopic surgery, duodenal atresia, one of the forms of CDO, also can be managed laparoscopically. However, use of a laparoscopic approach in these patients requires advanced experience in minimally invasive surgery and special equipment to perform such a demanding procedure in a limited operative space [3, 4]. Multiple operative techniques have been described, each with specific indications which will be discussed further in this chapter.

2. Background

2.1 Epidemiology

Congenital duodenal obstruction accounts for 50% of all intestinal atresia cases [1, 2]. The incidence of the disease varies from 1 in 5000 to 1 in 10,000 newborns [5–8]. Limited information is available about hereditary forms of CDO. Unlike other types of congenital intestinal obstruction, duodenal obstruction has a high association with other anomalies; such concomitant anomalies are reported in 38%

of patients with CDO [9, 10]. The most common associated disease reported is Down syndrome registered in 25–46% of cases [11–13]. Other associated anomalies include intestinal malrotation (54%), congenital cardiac anomalies (32–48%), esophageal atresia (9%), renal anomalies (5%), and anorectal malformations (7%) [14–20]. These anomalies can be part of VACTERL syndrome or isolated [13]. Spigland et al. reported that 12% of patients with duodenal atresia have an associated anomaly of the biliary tract, such as biliary atresia [21, 22]. Associated diseases tend to determine the postoperative course in patients with duodenal atresia. In patients with associated esophageal atresia or cardiac defects—most often a complete atrioventricular septal defect—high mortality rates have been reported [13, 23].

2.2 Embryology

Duodenal atresia results from an impaired development of the intestinal tube in early fetal life. The most common theory of duodenal atresia development was first described by Tandler et al. in 1900 and is based on failure of the solid core of duodenum to recanalize, also called the developmental arrest ("epithelial plug") theory [24–26]. Duodenal development is initiated at the beginning of the 4th gestational week from the distal part of the foregut and the proximal portion of the midgut. Between 5 and 6 weeks of intrauterine life, the lumen of the duodenum temporarily obliterates as a result of intensive proliferation of epithelial cells. Vacuolization, or degeneration of epithelial cells, occurs during the 11th week of gestation and leads to recanalization of the duodenal lumen [27]. Failure of this sequence in fetal development leads to duodenal atresia.

The vascular theory described in the middle of the twentieth century gained less popularity; however, it has been utilized for explanation of the types of duodenal atresia. According to this theory, a vascular accident leads to intrauterine bowel necrosis [28, 29].

Congenital duodenal obstruction can be caused by internal or external factors. Internal etiologies include intestinal atresia, stenosis, duodenal web, and intraluminal duodenal ("windsock") diverticulum. The latter is a fenestrated membrane in the second portion of the duodenum [30] prolapsing into the lumen of the distal duodenum. These internal forms of CDO are explained by the developmental arrest theory. External duodenal obstruction can result from annular pancreas, malrotation, or preduodenal portal vein. Most authors believe that the annular pancreas alone cannot cause CDO, but only accompanies atresia or stenosis of the intestinal lumen. Annular pancreas is an associated anomaly in patients with CDO [31]. However, the cause of CDO is failure of embryological development, not external compression of the duodenal lumen with the excessive pancreatic tissue [6]. This is well supported by the data obtained during the study of the embryologic development of the digestive tract.

CDO can be classified into pre- and postampullar. The latter is more common and is detected in 85% of affected newborns. In cases of complete obstruction, a dilated stomach and proximal duodenum are visualized, along with pyloric hypertrophy. The digestive tract distal to the obstruction is in a dormant state, excluding rare forms of type I atresia. In such cases, the membrane causing duodenal obstruction prolapses into the distal segment of the duodenum and its significant distention, obfuscating the true location of the obstruction. This anomaly is similar to a duodenal "windsock" diverticulum [32].

2.3 Classification

Congenital duodenal obstruction as described by Gray S. and Skandalakis J. is classified into three types [33]. Type I (the most common) is usually represented by an intraluminal membrane with a normal structure of the common muscular wall for the proximal and distal segments. Duodenal stenosis belongs to this type of CDO. Type II is characterized by a short fibrous cord between the blind segments of the duodenum with separate muscle layers. Type III (rarer) is the complete separation of duodenal segments. In this anatomical variation of duodenal obstruction, multiple atresias are often present and require intraoperative exploration of the small bowel distal to the duodenal obstruction, using a liquid test.

Partial congenital duodenal obstruction or congenital duodenal stenosis, most often fenestrated membrane, is a rare cause of high intestinal obstruction in neonates. The opening in the membrane allows for passage of food and can cause duodenal obstruction of various severity. Thus, the anomaly may not be detected immediately after birth; diagnosis can be delayed even into adulthood [34]. The symptoms can mimic other conditions, such as gastroesophageal reflux disease.

3. Diagnosis

In most cases, the diagnosis of duodenal obstruction can be established prenatally. Duodenal obstruction develops approximately by 12–14 weeks of fetal development, so there is no possibility of earlier detection of this anomaly. Ultrasound is used to define the "double bubble sign." These are two fluid levels, one in the distended stomach, and the other one in the duodenum (**Figure 1**). Polyhydramnios develops in pregnancies complicated by duodenal obstruction.

Postnatally, the diagnosis of duodenal obstruction is confirmed in an abdominal X-ray, showing the "double bubble" sign described above (**Figure 2**). Abdominal ultrasound is necessary to detect not only duodenal atresia (**Figure 3**), but also to find concomitant anomalies and rare forms of situs inversus. These findings can necessitate alternative port placement during laparoscopy.

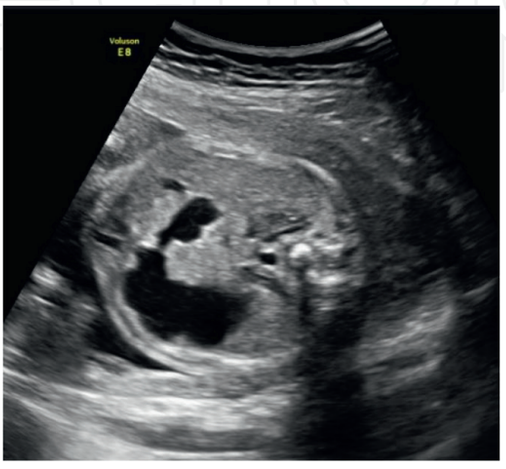

Figure 1.
"Double-bubble" sign of duodenal obstruction on a prenatal ultrasound.

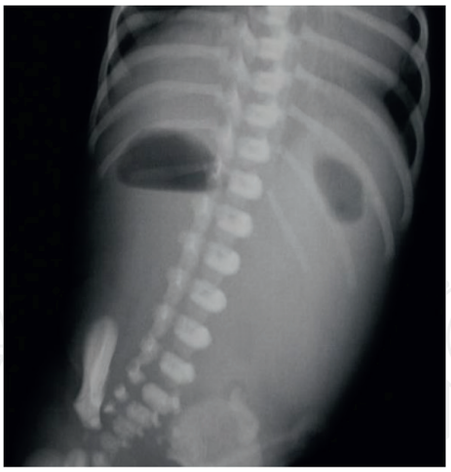

Figure 2.
Postnatal X-ray showing the "double-bubble" sign of duodenal obstruction.

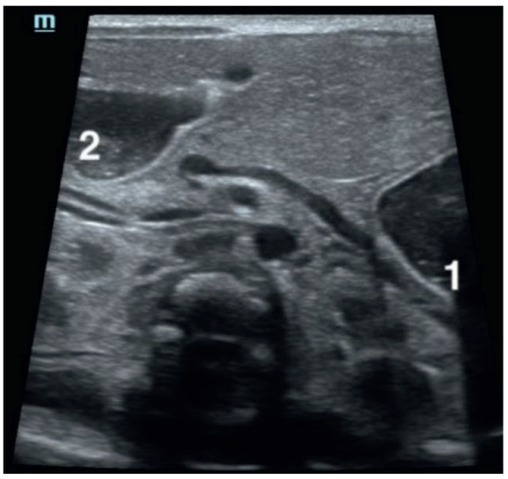

Figure 3.
Postnatal ultrasound showing the "double-bubble" sign of duodenal obstruction. 1 – Stomach, 2 – Duodenum.

4. Treatment

4.1 Surgical treatment options

Currently, the standard method of recanalizing the duodenal lumen is a diamond-shaped duodenal anastomosis, first reported by Kimura K. et al. [35]. Reconstruction is usually performed *via* right supraumbilical or umbilical incisions [10, 36–42]. The introduction of minimally invasive laparoscopic instruments, optical systems with small diameters, and high-resolution screens has expanded the potential of laparoscopy. These developments have increased the interest of pediatric surgeons in laparoscopy as a modality for reconstruction in patients with CDO [43]. Procedures considered complicated in the past have become both practical and effective [3, 4, 44, 45].

The first laparoscopic reconstruction for duodenal atresia was performed in 2001 by Bax N. [46]. In 2002, Gluer S. et al. described a case of laparoscopy reconstruction for annular pancreas and malrotation [47]; however, the initial experience of endoscopic duodenal anastomoses was overshadowed by a high number of conversions to

laparotomy and postoperative anastomotic leakage. Van der Zee D. even announced a temporary moratorium on performing minimally invasive interventions in patients with duodenal atresia due to excessive complications. He restricted the use of laparoscopic duodenal atresia repair to centers with extensive experience in pediatric laparoscopic surgery and intracorporeal suturing [3, 4, 48]. As more experience was gained, the operative technique was refined, and duodenal anastomosis could be performed with acceptable long-term results in this advanced center. Other studies also demonstrated the high efficacy and safety of laparoscopic surgery in the treatment of duodenal atresia [49–51]. Razumovsky A., one of the pioneers of neonatal minimally invasive surgery, has the greatest experience in performing laparoscopic duodenal anastomoses in Russia [52].

Several progressive surgical schools have presented modifications of laparoscopic duodenal anastomoses. Son T. et al. reported a side-to-side duodenal anastomosis [53]. Muensterer O. and Lacher M. showed that duodenojejunal side-to-side anastomosis can be performed to restore the lumen of the duodenum [54].

Laparoscopy allows for faster postoperative recovery and better cosmetic results. This knowledge promotes widespread use of laparoscopy in newborns and infants. However, laparoscopic duodenal anastomosis is considered the most demanding surgical procedure in pediatric surgery [48, 55]. Therefore, this procedure is restricted to be performed in advanced centers specializing in minimally invasive surgery in neonates [48]. Duodenal stenosis is most often located in the second (descending) portion of the duodenum [15]. Historically, such patients were treated surgically with laparotomy or laparoscopy. Despite the results of such treatment being satisfactory, these techniques are fraught with risks associated with the operation itself and general anesthesia, and do not show good cosmetical results [35, 46, 56]. In addition, the risk of adhesive intestinal obstruction in infants after laparotomy is approximately 6–14% [57] and is absent if the peritoneum is left intact, as, for example, with transoral access.

4.2 Endoluminal access

Endoluminal techniques for the treatment of duodenal obstruction based on principles of natural orifice transluminal endoscopic surgery (NOTES) were first introduced for adults in 1980 by Turnbull A. et al. using endoscopic biopsy forceps for membrane resection [58]. The first case of endoluminal surgery for congenital duodenal stenosis in children was reported by Okamatsu T. et al. In 1989, the author presented a successful case of endoscopic membranectomy in a 2-month-old child [59].

This technology involved the introduction of a small diameter occlusion catheter (5 Fr) into the opening of the fenestrated membrane, which, after inflating the balloon, stretches toward the stomach. The membrane spread on the catheter is dissected with a T-shaped incision using an electrode. Several other methods of duodenal web dissection have been reported, including the use of a sphincterotome [60, 61] and laser ablation [34, 62].

Several small case series demonstrated good results of partial CDO treatment using endoluminal balloon dilation [63–65]. The largest number of patients in the series included 6 children in whom balloon dilation was used as the first line of treatment [66]. The largest study of endoscopic membranotomy included 15 patients in whom dissection of the membrane was performed with a sphincterotome. One complication, perforation of the duodenum, was reported in this series [67].

There are several limitations to endoluminal methods in patients with partial CDO. The first one is a combination of duodenal stenosis with a more distal small bowel obstruction. Goring J. et al. described such a patient who subsequently underwent laparotomy and small bowel anastomosis [67]. The second limitation is annular pancreas, which reduces the chances of long-term success after endoluminal treatment [66]. Therefore, an abdominal MRI is recommended to assess the anatomy of the pancreas before transoral procedures [67]. If an annular pancreas is revealed on MRI, the use of endoluminal treatments is contraindicated.

4.3 Operative techniques

4.3.1 Laparoscopic Membranectomy and Heineke-Mikulicz Duodenoplasty

The patient is placed in the reverse trendelenburg position at the lower edge of the operating table to provide convenient access to the operating field for the surgeon (**Figure 4**). Veress needle access is performed and three laparoscopic ports are installed: (1) optical port in the left iliac region, (2) manipulation port in the left upper quadrant, (3) manipulation port in the right iliac region. The optical port is placed lower to allow for better visualization of the duodenal malformation (**Figure 5**). This port placement allows enough working space to perform duodenal anastomosis.

Low-pressure pneumoperitoneum with CO_2 is used: pressure of 8 mmHg and flow of 2 L/min. The first step is a transabdominal suture around the round ligament of the liver to fixate the liver to the abdominal wall. This maneuver improves visualization of the duodenum, especially its distal part. The hepatic flexure of the colon is mobilized using blunt or sharp dissection of its ligaments. A "no-touch" technique is preferred, capturing the surrounding tissues without crushing the intestinal walls with a clamp. Mobilization should be carried out widely enough to separate the transverse colon from the duodenum and allow for sufficient access to the duodenum. Importantly, as the duodenum bends around the pancreas it can be located either anteriorly or posteriorly, depending on the patient's unique anatomy. If the bulb of the duodenum is significantly stretched, a transparietal traction suture can be placed on the anterior

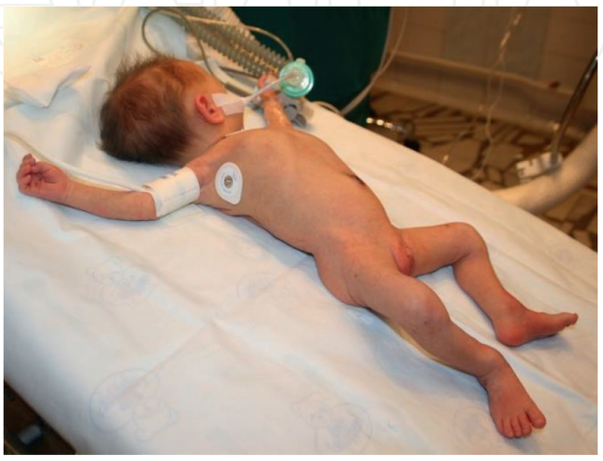

Figure 4.
Positioning of the patient for a laparoscopic reconstruction of duodenal atresia.

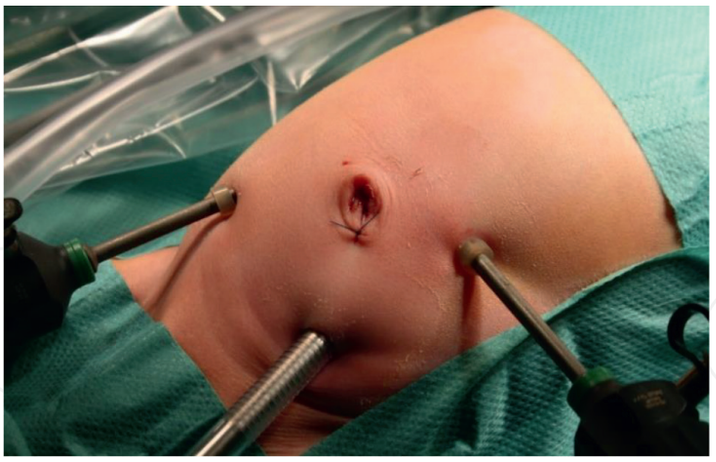

Figure 5.
Placement of the laparoscopic ports.

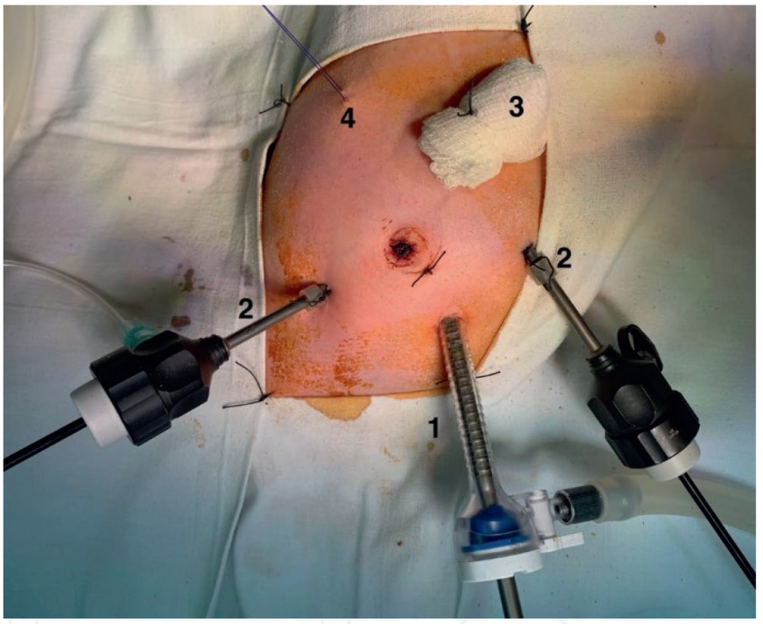

Figure 6.
Operative field with the optical port (1), trocars for manipulative instruments (2), traction suture over the round ligament of the liver (3), and traction suture on the proximal part of the duodenum (4).

wall of the bulb and brought out through the skin, allowing for mobilization of the distal portion of the duodenum (**Figure 6**). Lysis of adhesions can be performed with blunt dissection.

The location of the obstruction must be precisely defined. The obstruction can result from an annular pancreas, duodenal stenosis, or true duodenal atresia. At the time of mobilization of the distal portion of the duodenum, the gastrointestinal tract can be assessed for malrotation [68]. After that, the proximal and distal parts of the duodenum are identified and mobilized (**Figure 7**). Further steps depend on the type of duodenal anomaly.

The findings determining the type of reconstruction are (a) membranous, type I, or (b) other forms of obstruction with separation of the muscle membranes of disconnected segments, including duodenal atresia with (type II) or without (type III)

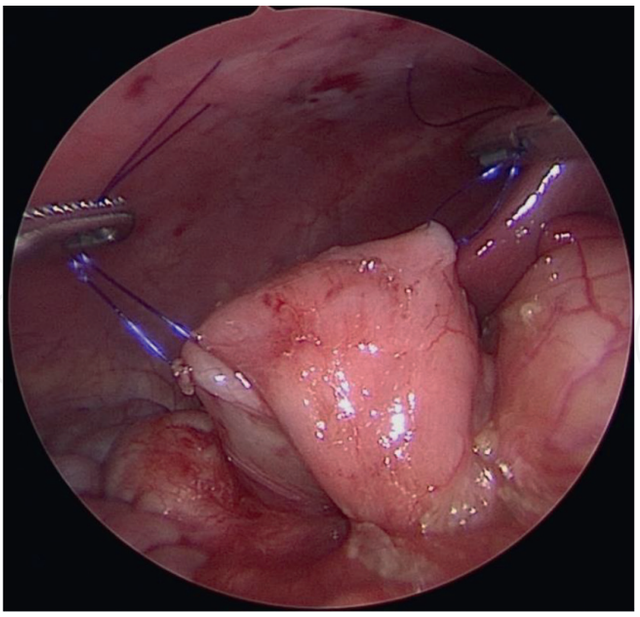

Figure 7.
Duodenal atresia type 1.

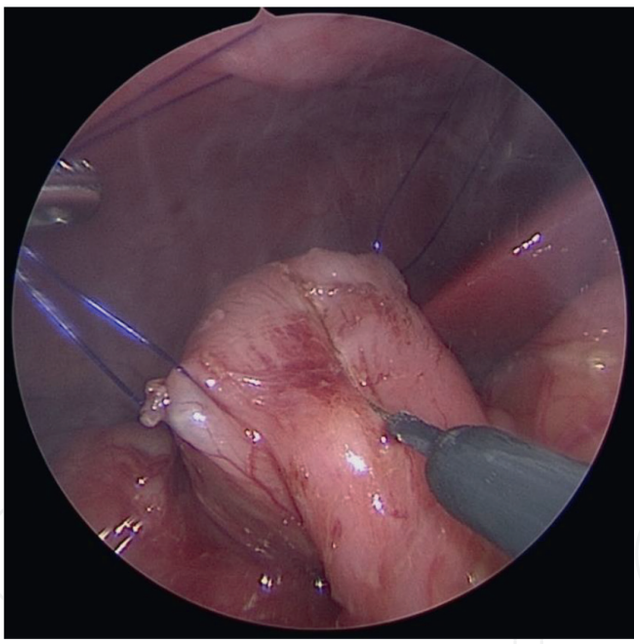

Figure 8.
Longitudinal duodenotomy over the site of obstruction.

a fibrous cord and an annular pancreas. Membranous forms of duodenal obstruction are repaired using a longitudinal duodenotomy over the site of obstruction with incision of the proximal and distal portions of the duodenum (**Figures 8** and **9**). Then, the membrane is captured with a delicate atraumatic clamp and excised circularly with coagulation micro-scissors (**Figures 10** and **11**). Particular care is necessary for dissecting the duodenum at the ampulla of Vater [69]. The final step of the operation is transverse suturing of the duodenal lumen with interrupted absorbable sutures (**Figures 12** and **13**).

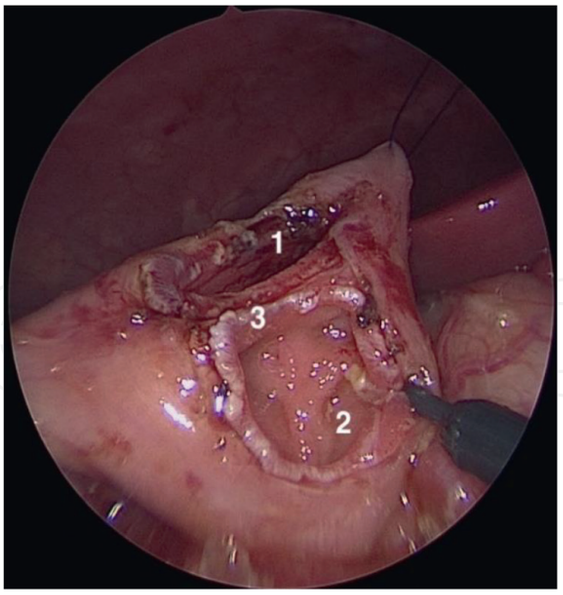

Figure 9.
Duodenum after its excision. 1 - proximal part, 2 – Distal part, 3 – Duodenal membrane.

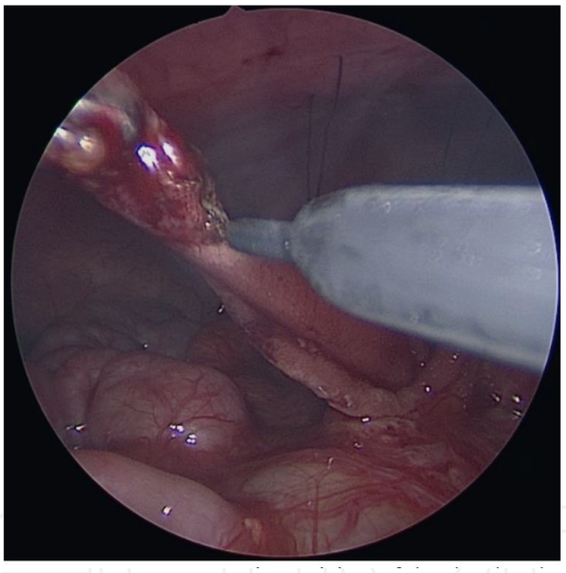

Figure 10.
Laparoscopic excision of the duodenal web.

4.3.2 Laparoscopic Kimura anastomosis

In patients with duodenal atresia type II and type III, a diamond-shaped Kimura anastomosis is performed (**Figure 14**). The technique of laparoscopic diamond-shaped anastomosis does not differ from the open procedure (**Figure 15**) [39, 70]. A transverse duodenotomy of the dilated proximal part of the duodenum and a longitudinal incision of the distal collapsed segment are performed using a needle or hook-shaped electrode (**Figures 16** and **17**). A transparietal traction suture is placed on the proximal portion of the duodenum, which is brought to the anterior abdominal wall to allow for fixation of the bowel to allow for duodenal anastomosis creation.

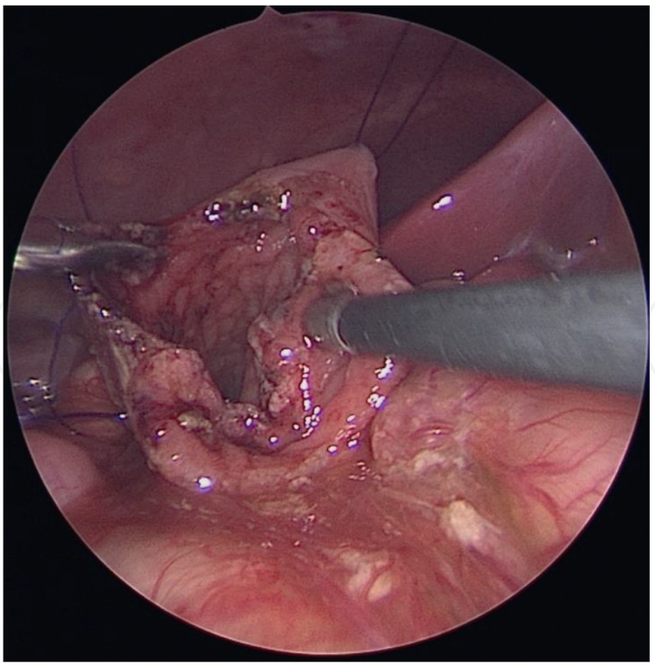

Figure 11.
Lumen of the duodenum after resection of the duodenal web.

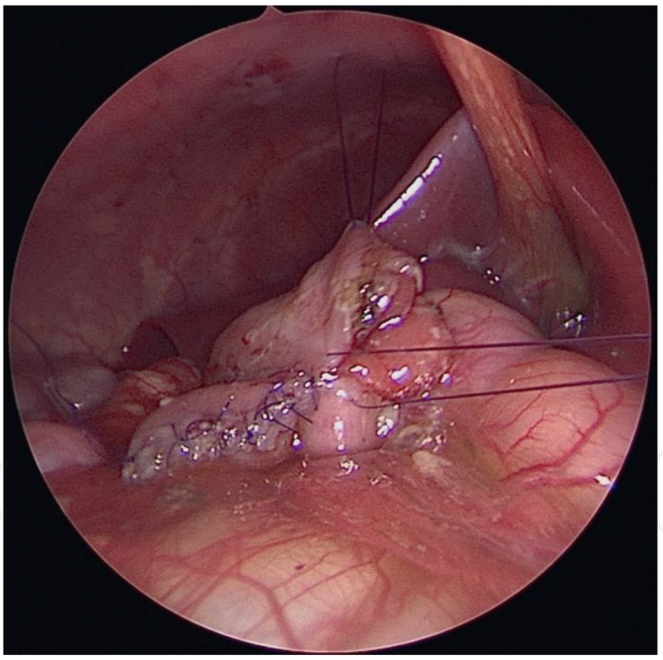

Figure 12.
Suturing of the longitudinal duodenotomy in a transverse fashion.

The technique of laparoscopic Kimura anastomosis involves placement of separate sutures (PDSII 6/0), on the posterior (**Figure 18**) and then the anterior wall of the duodenal junction (**Figure 19**). To achieve the diamond shape, the first suture is placed between the right point of the transverse duodenotomy and the middle of the right part of the longitudinal duodenotomy. Subsequent stitches are placed bilaterally to the right and left of this initial suture so that the final stitches of the posterior wall of the anastomosis are placed between the right and left ends of the transverse

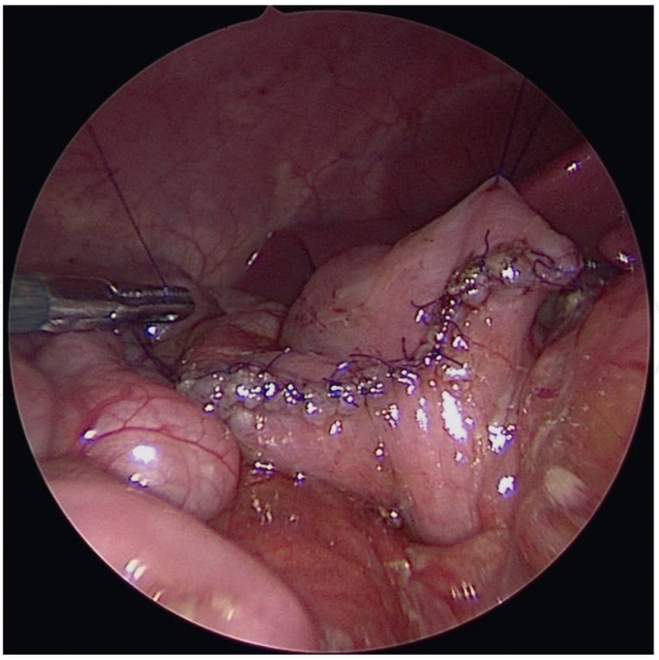

Figure 13.
Duodenal anastomosis after resection of the duodenal web.

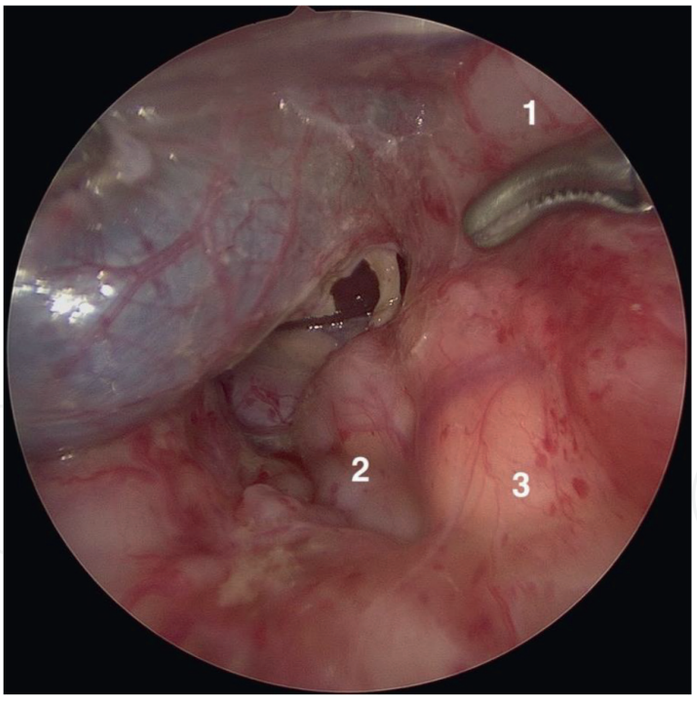

Figure 14.
Duodenum type 3. 1 – Proximal segment, 2 – Distal segment, 3 – Head of the pancreas.

incision and the middle of the longitudinal incision on both sides. Thus, the lower triangle of the future diamond-shaped anastomosis is formed. The anterior wall of the anastomosis is created in a similar way, connecting initially the central points of the incisions, and then the peripheral ones. The result of these complicated actions on small objects in a limited field for manipulation is a wide duodenoduodenal anastomosis. Van der Zee D. places the second suture between the left corner of the

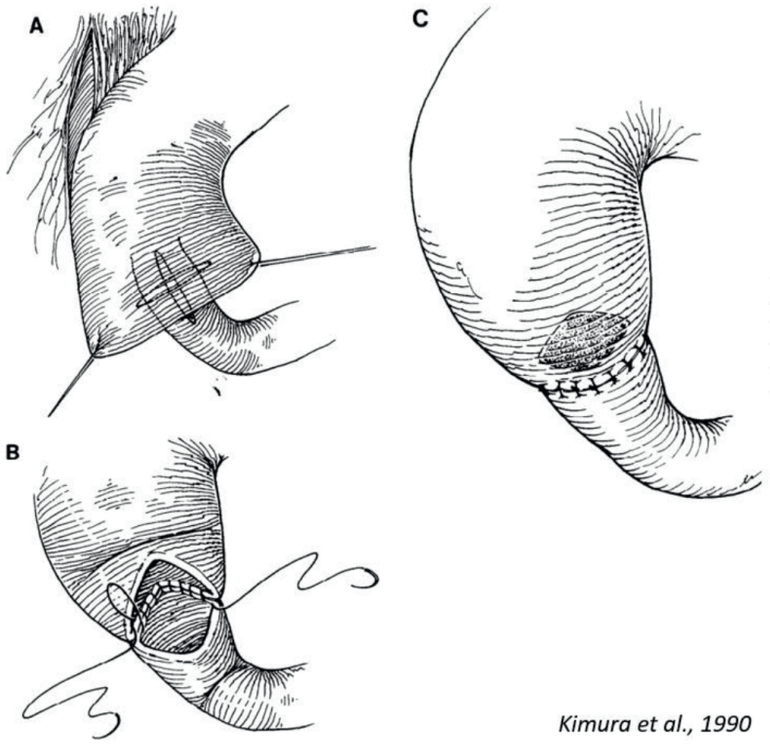

Figure 15.
A diamond-shaped anastomosis [37].

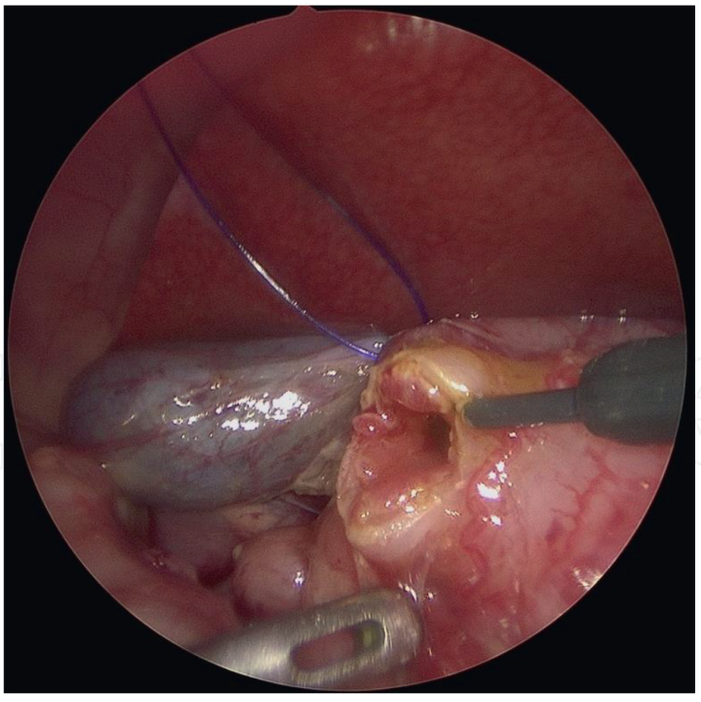

Figure 16.
Excision of the proximal segment of the duodenum.

transverse duodenotomy and the middle of the left edge of the longitudinal duodenotomy (outside-in, inside-out). After tying this stitch, the needle is placed inside the intestinal lumen and a continuous suture is placed on the posterior wall from inside

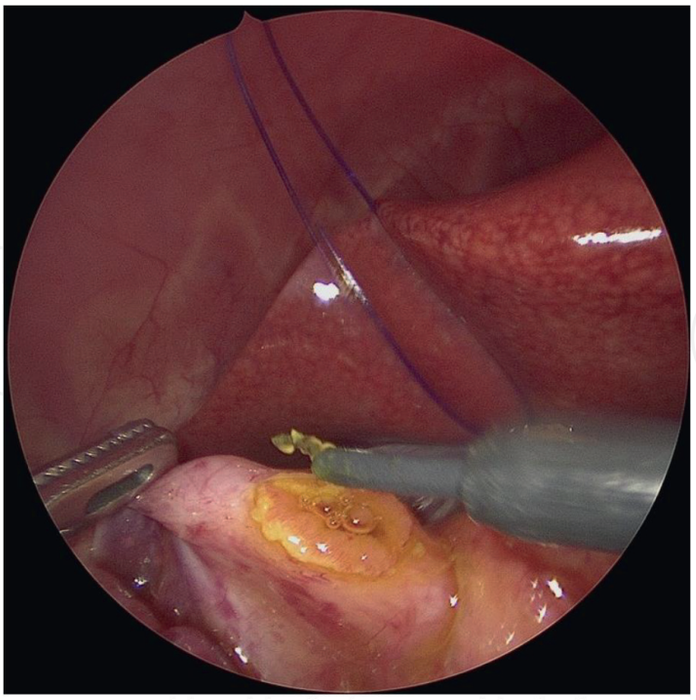

Figure 17.
Excision of the distal segment of the duodenum.

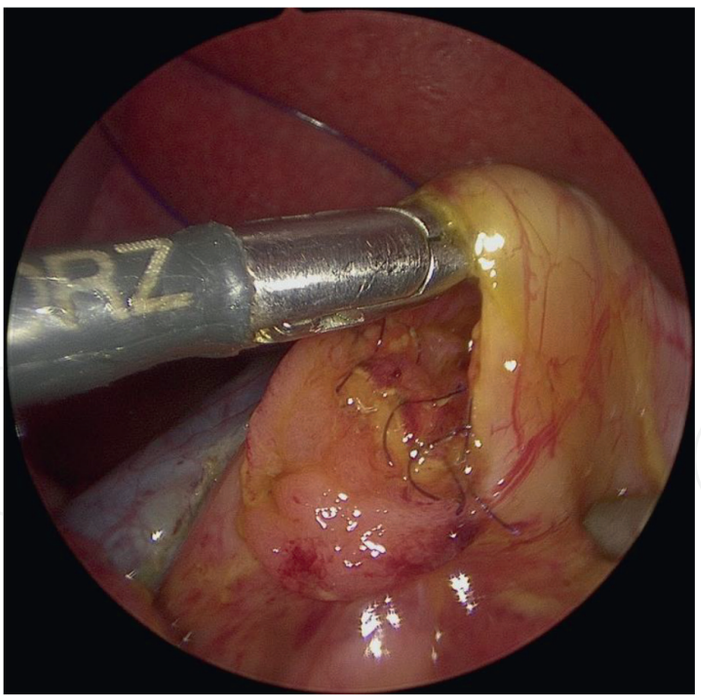

Figure 18.
Duodenal anastomosis described by Kimura K. et al. [33]. Creation of the posterior wall of the anastomosis.

the lumen. The posterior wall of the anastomosis can be easily visualized for a future continuous suture by applying traction to the left stitch.

At the right end of the suture line on the posterior wall, the needle is brought out and tied with the short end of the first suture. The same thread can be used to

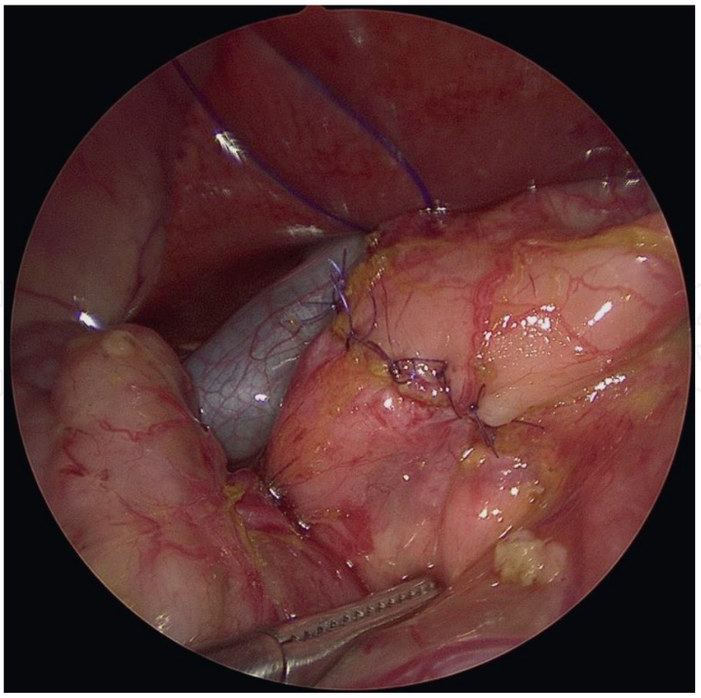

Figure 19.
Duodenal anastomosis described by Kimura K. et al. [33]. View at the end of the procedure.

perform a continuous suture on the anterior wall of the anastomosis. In cases of an annular pancreas, the anastomosis is created in the form of a "bridge" over the area of ectopic glandular tissue-duodenal "bypass" (**Figures 20** and **21**). It is important to make sure that this is exactly the horizontal part of the duodenum, not the loop of the jejunum because this can lead to the creation of a blind loop. At the end of the operation, no feeding tube is passed through the area of the anastomosis as there is no need to inspect for other membranes or obstruction sites, given that these are extremely rare (less than 1%) [71]. The anesthesiologist performs the white test (injection of a fat emulsion solution for parenteral nutrition into a pre-installed gastric tube). This maneuver confirms there is no anastomotic leak or intestinal obstruction in the distal part of the small bowel.

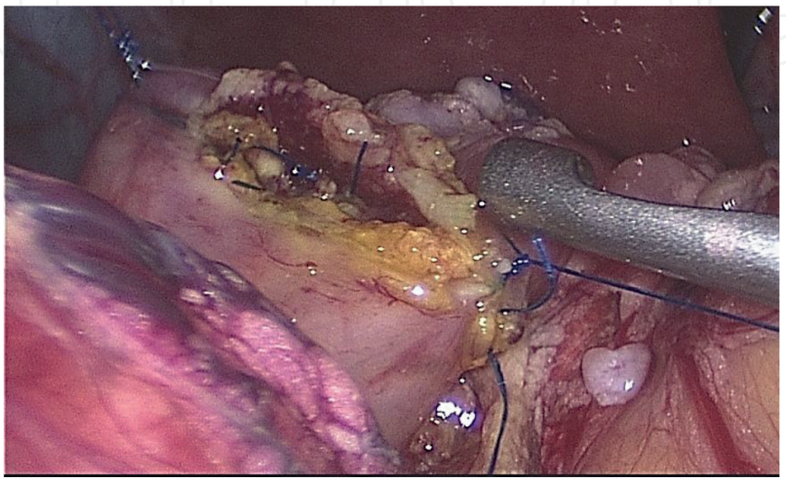

Figure 20.
Suturing of the anterior wall of a duodenal anastomosis in patients with an annular pancreas.

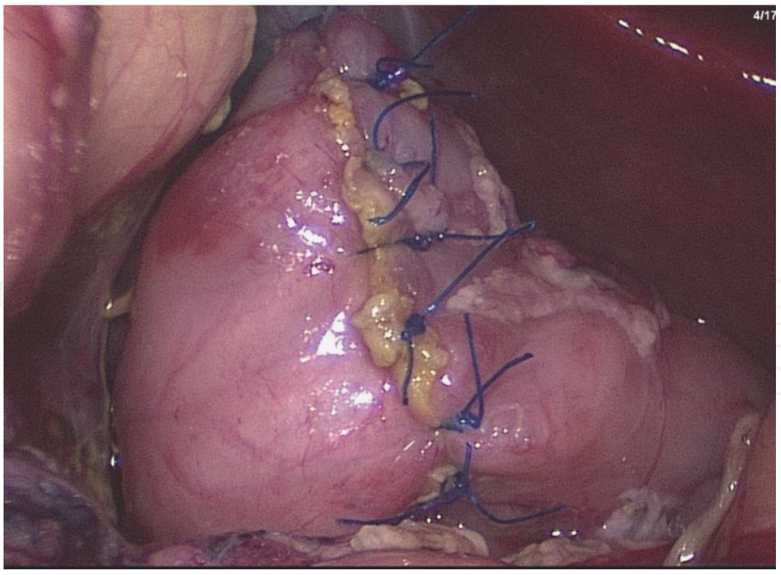

Figure 21.
Duodenal "pathway" anastomosis in a patient with an annular pancreas.

4.3.3 Transoral endoscopic Membranotomy and balloon dilation

The procedure is performed under general endotracheal anesthesia using a single-channel video gastroscope with a 3.2-mm instrument channel. Before the operation, the remnants of formula and saliva are removed from the stomach with irrigation and aspiration. After passing the pylorus, a dilated superior (first) segment of the duodenum is visualized blindly ending in a membrane with an opening is its center (**Figure 22**). When using the method of balloon dilation, a CRE Fixed Wire balloon catheter (Boston Scientific Corporation, USA), diameter of 10–11-12 mm and inflation pressure of 3–5-8 atm, is inserted into the opening of the duodenal membrane (**Figures 23** and **24**). After dilation of the lumen, the endoscope is advanced to the distal portion of the duodenum. Endoscopic membranotomy can also be performed by advancing of a double-lumen

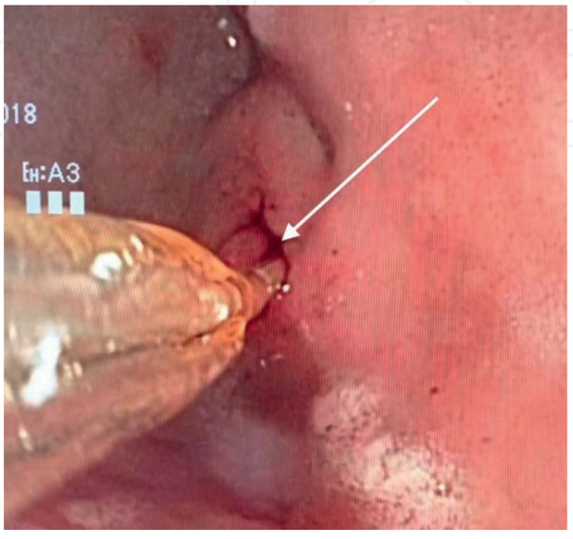

Figure 22.
Opening in the duodenal web (arrow) with the balloon catheter CRE fixed wire (Boston Scientific Corporation, USA) inserted.

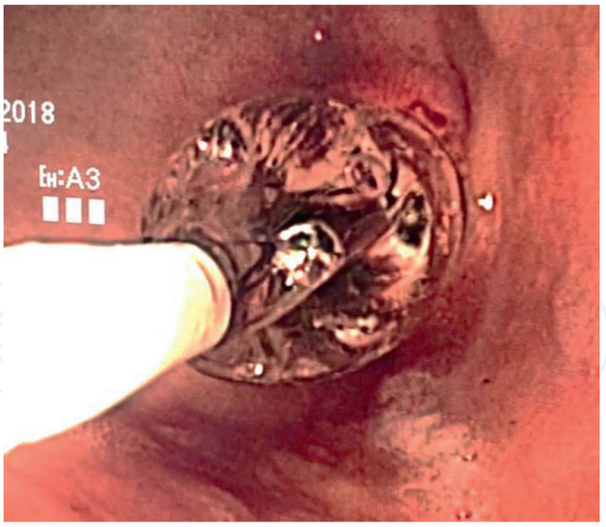

Figure 23.
Balloon dilation of duodenal stenosis with a catheter CRE fixed wire (Boston Scientific Corporation, USA).

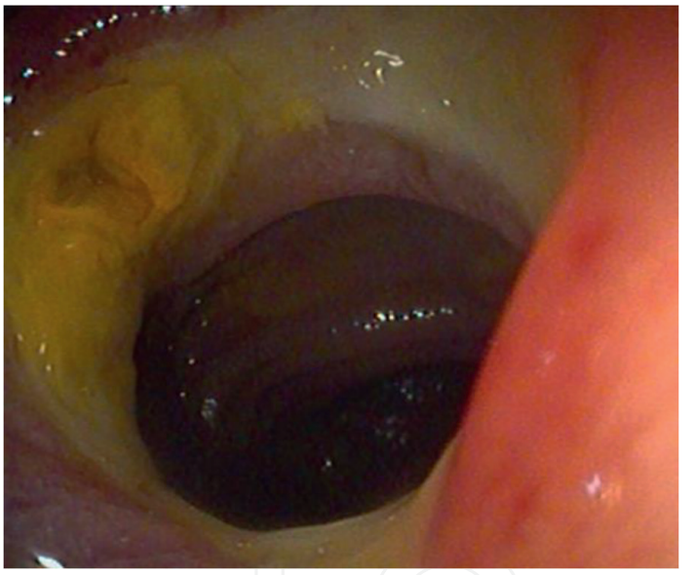

Figure 24.
Area of duodenal stenosis on postoperative day 5 after balloon dilation.

catheter designed for sphincterotomy - Minitome double lumen sphincterotome (Cook Medical, USA) with a length of 200 cm—to the location of duodenal stenosis. After the device is activated, mixed current is supplied to it from the electrosurgical unit. Bleeding from the edges of the membrane can be stopped using a soft coagulation mode. After destruction of the membrane, the endoscope is advanced into the distal portion of the duodenum.

Postoperatively, patients are observed in the intensive and critical care unit until their respiratory and circulatory functions are stabilized. Enteral nutrition is initiated by the end of the first postoperative, as the lumen of the bowel was not opened. Diet advancement is based on tolerance to increasing amounts of nutrition, with gradual increase of nutrition volumes within the first two postoperative days.

5. Conclusion

Laparoscopy, as a less aggressive surgical technique, offers an alternative approach to the treatment of congenital duodenal obstruction, allowing for minimally invasive access and early recovery after the surgery. Since the first report of a successful laparoscopic anastomosis in a patient with duodenal atresia, laparoscopic reconstruction was expected to become a golden standard of congenital duodenal obstruction treatment. However, its complexity and time consumption prevented this technique from being widely used among pediatric surgical centers compared to an open repair. Laparoscopic reconstruction remains the prerogative only for advanced pediatric surgery centers specializing in minimally invasive surgery in neonates. Transoral techniques based on flexible endoscopy have a special place in minimally invasive treatment of partial duodenal obstruction and allow for expansion of the narrow site using a balloon or a sphincterotome with no incision required.

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