Langenbeck's Archives of Surgery (2022) 407:835–844
https://doi.org/10.1007/s00423-022-02460-5

HOW-I-DO-IT ARTICLES

Surgical strategies for duodenal gastrointestinal stromal tumors

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Received: 2 August 2021 / Accepted: 2 February 2022 / Published online: 18 February 2022

Abstract

Purpose Duodenal gastrointestinal stromal tumors (GISTs) are rare mesenchymal tumors of the gastrointestinal tract. For localized or potentially resectable GISTs, surgery is the first choice. But the important and complex anatomical structure adjacent to the duodenum makes surgical management for duodenal GISTs challenging and few comprehensive surgical strategies have been described. This study aims to provide new comprehensive surgical strategies for duodenal GISTs by summarizing the surgical approaches and outcomes of duodenal GISTs in different locations in our center in the past 11 years.

Methods Information from patients who underwent surgical resection for duodenal GISTs at our facility during the past 11 years was retrospectively analyzed.

Results Ninety-two patients have received surgical procedures in the facility. Twenty-three, 31, 3, and 35 patients underwent wedge resection, segmental resection, pancreatic head-preserving duodenectomy, and pancreaticoduodenectomy, respectively. The mean operative times were 212.6 (150–270), 260 (180–370), 323 (300–350), and 354.9 (290–490) min; the mean blood loss was 226.1 (100–400), 303.2 (100–600), 500 (400–600), and 582.9 (200–1300) ml, respectively. R0 margins were obtained in 21, 29, 3, and 32 patients, respectively.

Conclusions For duodenal GISTs without invasion of the ampulla of Vater or the pancreatic head, a limited resection (such as wedge resection, segmental resection, or pancreatic head-preserving duodenectomy) is feasible. For duodenal GISTs with an invasion of the ampulla of Vater or the pancreatic head, a pancreaticoduodenectomy is still necessary.

Keywords Duodenum · Gastrointestinal stromal tumors · Surgery · Pancreaticoduodenectomy

Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract with diverse biological behaviors. Most GISTs have c-kit or platelet-derived growth factor receptor alpha gene activating mutations [1, 2]. They occur most commonly in the stomach (60%) and small intestine (30%), but only 4–5% are located in the duodenum [3]. For localized or potentially resectable GISTs, surgery is the first choice. The surgical procedure should be aimed at the removal of an intact tumor with a negative histological margin, and complex multiorgan resection should be avoided to minimize the surgical complications [4].

The important and complex anatomical structure adjacent to the duodenum, such as the head of the pancreas, ampulla of Vater, and common bile duct, makes surgical management for duodenal GISTs challenging, and the removal of tumors of the duodenum often involves the pancreatic head. Currently, a variety of surgical approaches, such as segmental or wedge-shaped duodenectomy and pancreaticoduodenectomy (PD), have been described for duodenal GISTs [5, 6]. The goal to achieve histologically negative margins, simplify the surgical procedure, and preserve the organ as much as possible requires a balance between PD and limited resection due to the limited intramural extension of GISTs, but few comprehensive surgical strategies have been described. The purpose of this study is to provide new comprehensive surgical strategies for the surgical management for duodenal GISTs

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by summarizing the surgical approaches and outcomes of duodenal GISTs in different locations in our center in the past 11 years.

Material and methods

This study was approved by the local ethical review board of the West China Hospital and then the informed consent for participation was obtained.

Patients diagnosed with duodenal GISTs and underwent surgical resection at West China Hospital, Sichuan University, between January 2010 and May 2021 were retrieved from our database, and the cases that underwent endoscopic resection were excluded. The diagnosis was based on medical history, blood test, upper digestive endoscopic ultrasonography, contrast-enhanced CT, and MRI scan. A biopsy was performed only if it was necessary to confirm the diagnosis of primary GIST to begin preoperative treatment [4]. The surgical approach was determined after the presence of adjacent structural invasion, and distant metastasis was assessed and the resectability was evaluated.

The following variables were studied: patient demographics, primary tumor location, tumor size, laboratory results, (neo)adjuvant tyrosine kinase inhibitor (TKI) use, surgical approach, operative time, estimated blood loss, volume of blood transfusion, intraoperative and postoperative complications, pathological results, and length of hospital stay. Postoperative complications were graded by Clavien-Dindo criteria [7], and postoperative pancreatic fistula (POPF) and delayed gastric empties (DGE) were graded according to the definitions of the international study group of pancreatic surgery (ISGPS) [8–10]. The risk classification is based on the National Institutes of Health (NIH) criteria [11].

Data are reported as averages, ranges and percentages. Overall survival (OS) and recurrence-free survival (RFS) were estimated by the Kaplan–Meier method and compared with log rank test. Cox proportional hazard regression analysis was used to estimate the hazard ratio (HR) and its 95% confidence interval (CI) for limited resection group vs PD group comparison. All data were analyzed using SPSS software (version 26.0).

Surgical technique

Since the goal is to achieve the removal of an intact tumor with a negative histological margin with no routine lymph node dissection is required, the exact surgical procedure is closely related to the location and size of the tumor, the involvement of surrounding structures, and the distance from the ampulla of Vater. The surgical procedures are briefly summarized as follows (Table 1).

For small tumors in all the portions of the duodenum, if they were located away from the ampulla, a wedge resection of the duodenum was performed if possible. The incision margin was about 1–1.5 cm from the tumor margin. If feasible, a primary closure was performed, if not, a side-to-side duodenojejunostomy (Roux-en-Y manner) was performed. Otherwise, other surgical approaches were adopted.

For the tumors in the first portion of the duodenum, a segmental resection of the first portion (maybe combined with distal gastrectomy if the pylorus was involved) was usually performed. First, the hepatic flexure of the colon was mobilized and a Kocher's maneuver was performed to expose and mobilize the second portion of the duodenum. The gastrocolic ligament and gastrohepatic ligament were dissected to expose the posterior side of the stomach. The distal stomach and duodenal bulb were then separated from their posterior attachments. If the pylorus was involved, the right gastroepiploic vessels and right gastric vessels were ligated. After that, the duodenal bulb was disconnected from the gastric antrum if the distal stomach was not involved; otherwise, the gastric antrum was disconnected from the gastric body with a stapler device. The first portion of the duodenum (and the gastric antrum if it was to be resected) was gradually separated from the pancreatic head, while careful operation was carried out to avoid damage to the common bile duct and the ampulla of Vater. The duodenum was then divided between the first and second portions. The second portion of the duodenum was anastomosed with the gastric antrum end-to-end (Fig. 1A, B). If the gastric antrum was removed, the jejunum 15 cm distal from the ligament of Treitz was anastomosed with the gastric body side-to-side (Billroth's II).

Table 1 The surgical strategies

Location	The distance to the ampulla	Surgical approach
The first portion		Wedge resection or segmental resection
The second portion	> 1.5 cm	Wedge resection or segmental resection
	< 1.5 cm, without invasion of ampulla and pancreatic head	PHPD
	< 1.5 cm, with invasion of the ampulla or the pancreatic head	PD
The third and fourth portions		Wedge resection or segmental resection

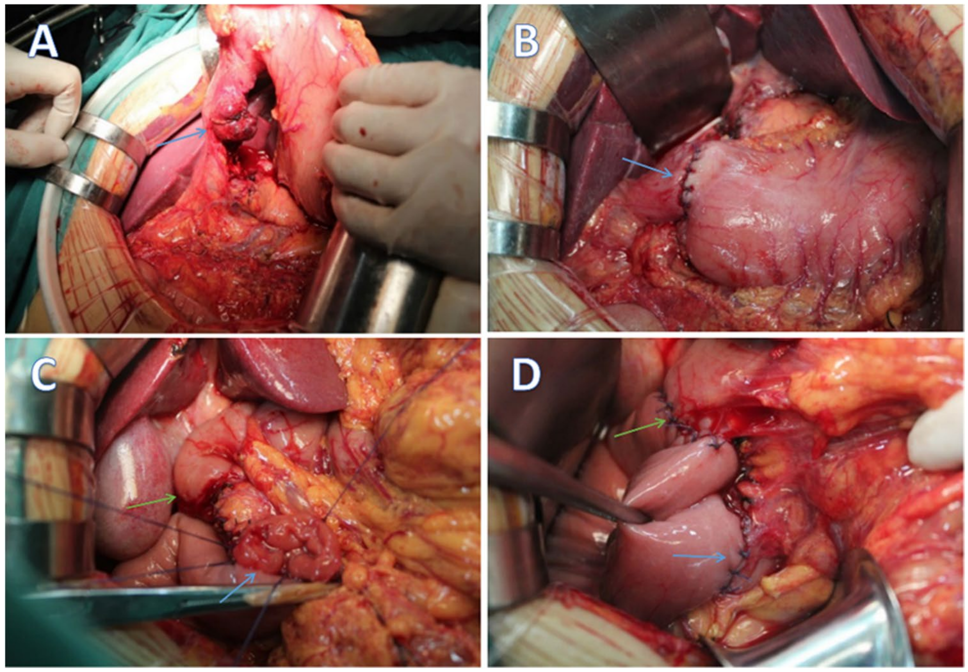

Fig. 1 **A** The blue arrow shows the tumor in the first portion of the duodenum. **B** The second portion of the duodenum was anastomosed with the gastric antrum end-to-end. **C** The green and blue arrows show the proximal and distal ends of the duodenum, respectively, after the proximal part of the second portion was removed. **D** The green and blue arrows show the proximal and distal parts of the duodenum which were anastomosed with jejunum, respectively

For tumors in the proximal part of the second portion of the duodenum which were located more than 1.5 cm away from the ampulla of Vater, a segmental resection of the second portion was performed. The method of mobilization and exposure of the duodenum was the same as above. After identifying the duodenal bulb, the duodenum was divided between the first and the second portions. A plane was developed between the second portion of the duodenum and the pancreatic head to separate them. The procedure needs to be carefully done to avoid injury to the pancreaticoduodenal vascular, the common bile duct, and the ampulla of Vater. The distal second portion of the duodenum was divided when the incision margin was more than 1 cm from the tumor margin, and usually reached the margin of the ampulla. The proximal and distal parts of the duodenum were anastomosed with the jejunum end-to-side and end-to-end respectively, via a Roux-en-Y manner (Fig. 1C, D, Fig. 4A–C).

For tumors in the second portion of the duodenum which were located close to the ampulla of Vater (< 1.5 cm), but without invasion of the ampulla and pancreatic head, pancreatic head-preserving duodenectomy (PHPD) was performed. First, the hepatic flexure of the colon was mobilized; then, a Kocher's maneuver was performed to expose and mobilize the pancreatic head and the second and third portions of the duodenum. The distal stomach and duodenal bulb were mobilized the same way as above, and the third portion was mobilized inferiorly and posteriorly. After identifying the pylorus, the proximal duodenum was divided 1 cm past the pylorus to save it, and the distal duodenum was divided between the second and third portions with a stapler device. A plane was developed between the first and second portions and the pancreatic head to separate the duodenal-pancreatic head complex. The first and the second portions were then dissected off the pancreatic head gradually by using ultrasonic shears. To avoid injury to the pancreaticoduodenal vascular system, we prefer to ligate these branch vessels close to the bowel wall. This process needs to be done very carefully, because as we get closer to the ampullary region, the attachment between the duodenum and the pancreatic head becomes denser. It is preferable to open the duodenum and place a catheter through the ampulla of Vater if it is hard to identify the common bile duct. The accessory pancreatic duct was ligated if it was identified. Then, the pancreaticobiliary duct was transected after the first and second portions of the duodenum were completely separated from the pancreatic head. The digestive tract was reconstructed via a Roux-en-Y manner. An end-to-side pancreaticojejunostomy was performed about 5 cm distal from the jejunum stump, using the same method as we did in PD. If the main pancreatic duct and the common bile duct did not have a common channel, a septotomy was performed to create one. First, the pancreatic capsule, including a portion of pancreatic parenchyma, was anastomosed to the jejunal seromuscular layer to form the posterior layer. After that, a small hole was made in the jejunum, and a 5F catheter was inserted through the hole, passing through the jejunal lumen and the anastomotic stoma into the pancreatic duct to drain the pancreatic juice and act as a stent. Then, a duct-to-mucosa pancreaticojejunostomy was performed. All the jejunal layers were anastomosed to the pancreaticobiliary duct complex, with a running suture. Finally, the anterior layer of the pancreaticojejunostomy was performed in a manner similar to the posterior layer. The pylorus was anastomosed with the jejunum end-to-side 45–60 cm distal from the pancreaticojejunostomy (Fig. 2, Fig. 4D–F).

For tumors located close to the ampulla (< 1.5 cm) and/or with an invasion of the pancreas or ampulla, a PD was performed. Since PD is a routine surgical procedure, it will not be described here.

For tumors in the distal part of the second portion of the duodenum that were more than 1.5 cm from the ampulla, a segmental resection of the distal part of the second portion was performed. The method of mobilization and exposure of the duodenum was the same as above. After resection, an end-to-end duodenal anastomosis was performed if possible (Fig. 3A, B); otherwise, the distal duodenum stump was closed and the proximal duodenum stump was anastomosed with the jejunum end-to-side via a Roux-en-Y manner.

For tumors in the third portion of the duodenum, a segmental resection was performed. The method of mobilization and exposure of the second portion and third portion was the same as above. For tumors in the fourth portion, a segmental resection was also performed. First, the transverse colon was pulled upward to expose the beginning of the jejunum. The ligament of Treitz was taken down, the fourth portion was mobilized inferiorly and posteriorly, and the proximal jejunum was then divided with a stapler device. The fourth portion was dissected off the pancreas caudad to cephalad gradually by using ultrasonic shears. Once the fourth portion was fully mobilized, it was divided from the third portion. After resection, an end-to-end duodenojejunostomy was performed if possible; otherwise, a side-to-side duodenojejunostomy was performed via a Roux-en-Y (Fig. 3C–F).

Results

The demographic characteristics, tumor characteristics, (neo)adjuvant tyrosine kinase inhibitor use, and surgical outcomes details are shown in Table 2. In the past 11 years, 92 patients with duodenal GISTs have received surgical procedures in our department. Twenty-three patients underwent wedge resection, including 9 males and 14 females, with a mean age of 49.8 (13–71) years. Segmental resection was performed in 31 patients, including 15 males and 16 females, with a mean age of 48.2 (26–74) years. PHPD was performed in 1 male and 2 female patients, with a mean age of 50.3 (38–65) years. Thirty-five patients underwent PD, including 19 males and 16 females, with a mean age of 51.1 (34–71) years.

Of the patients who underwent wedge resection, the mean tumor size is 2.3 (0.8–3.0) cm; 9 tumors were located in the first portion, 8 tumors in the second portion, and 6 tumors in the third portion. None of the patients received neoadjuvant TKI, and 4 patients (17.4%) received postoperative adjuvant TKI. The mean operative time was 212.6 (150–270) min and the mean blood loss was 226.1 (100–400) ml. Biochemical leakage was observed in 1 case (4.3%). Grade A DGE was observed in 2 cases (8.7%) and grade B in 1 case (4.3%). No complications greater than Clavien-Dindo grade II were observed. The mean postoperative hospital stay was 9.4 (5–25) days. R0 margin was achieved in 21 cases (91.3%)

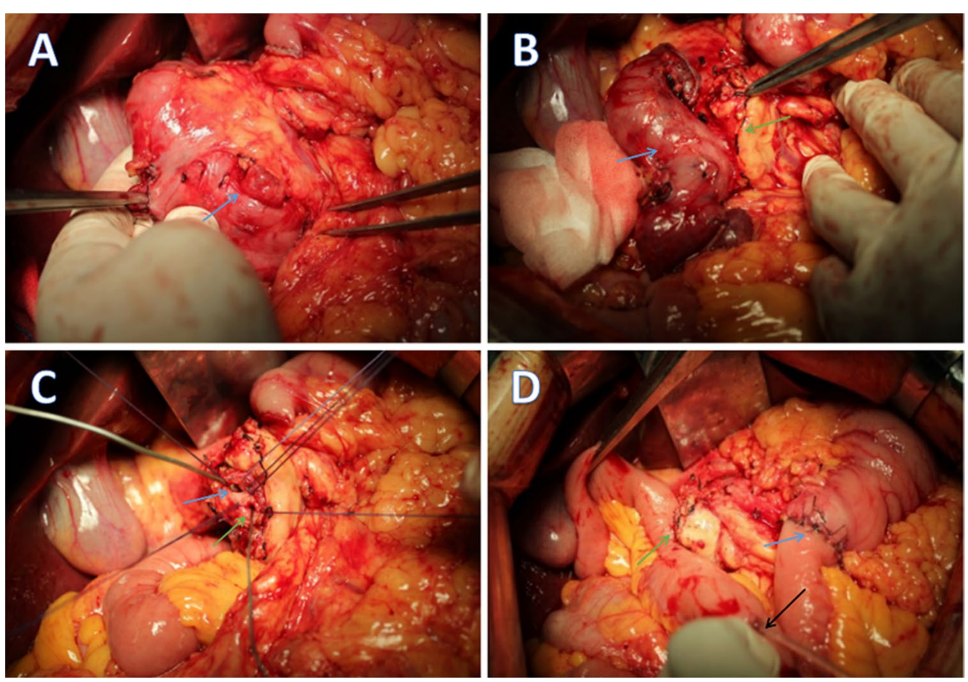

Fig. 2 PHPD. **A** The blue arrow shows the periampullary tumor that did not involve the pancreatic head. **B** The blue arrow shows the first and second portions of the duodenum which were completely separated from the pancreatic head; the pancreatic head is shown by the green arrow. **C** The pancreaticobiliary duct: the blue and green arrows show the common bile duct and the main pancreatic duct, respectively. **D** The blue and green arrows show the gastrojejunostomy and the pancreaticojejunostomy, respectively; the black arrow shows the drainage tube of the pancreatic duct

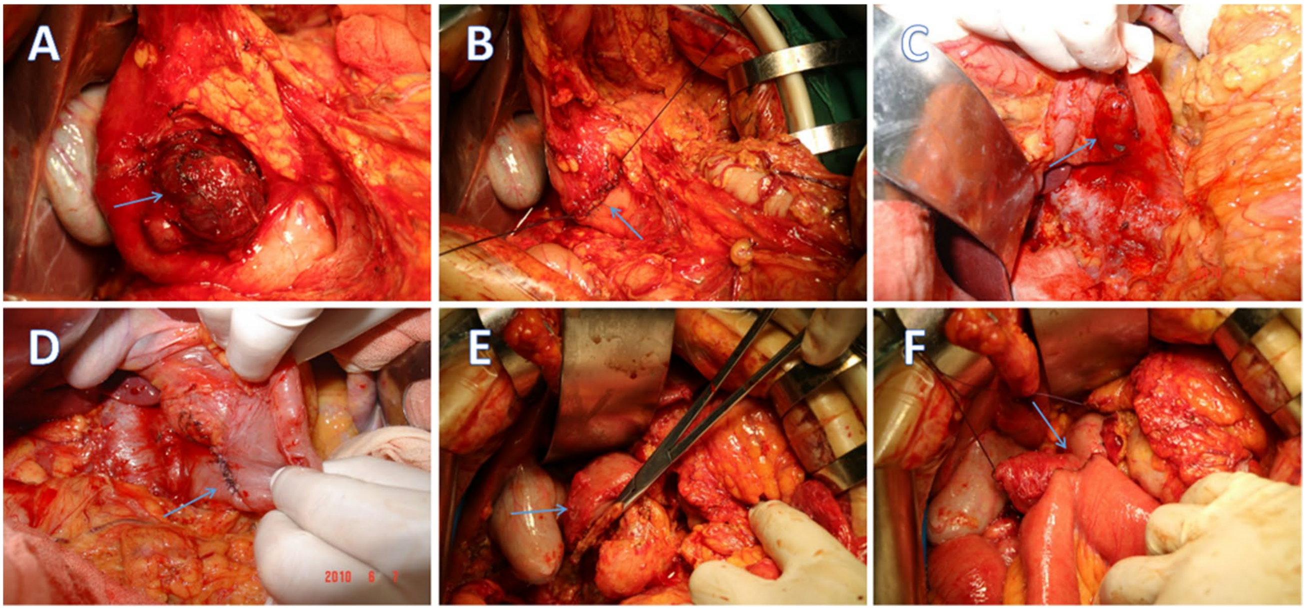

Fig. 3 **A** The blue arrow shows the tumor in the distal part of the second portion of the duodenum without involvement of the ampulla and the pancreas. **B** The blue arrow shows the end-to-end duodenal anastomosis after resection. **C** The blue arrow shows the tumor in the third portion of the duodenum. **D** The blue arrow shows the end-to-end duodenal anastomosis after resection. **E** The blue arrow shows the third portion of the duodenum after the fourth portion was resected. **F** The blue arrow shows the side-to-side duodenojejunostomy

and R1 margin in 2 cases (8.7%). Three cases (13.0%) were very low risk, 8 cases (34.8%) were low risk, 6 cases (26.1%) were intermediate risk, and 6 cases (26.1%) were high risk according to the NIH risk classification.

Of the patients who underwent segmental resection, the mean tumor size is 4.5 (1.7–14.0) cm; 10 had tumors in the first portion, 12 in the second portion, 4 in the third portion, and 5 in the fourth portion. One patient (3.2%) who received neoadjuvant TKI had a 14-cm-diameter tumor in the fourth portion suspected of involving SMA/SMV, but the final surgical margin was R1. Postoperative adjuvant TKI was performed in 9 cases (29%). The mean operative time was 260 (180–370) min and the mean blood loss was 303.2 (100–600) ml. One case of biochemical leakage (3.2%), 4 cases of grade A DGE (12.9%), 2 cases of grade B DGE (6.5%), 2 cases of grade C DGE (6.5%), and 1 case of postoperative hemorrhage (3.2%) were observed, which required a second laparotomy to stop bleeding, and was accompanied by pulmonary infection and respiratory dysfunction (Clavien-Dindo grade IV). The mean postoperative hospital stay was 14.9 (5–102) days. R0 margin was achieved in 29 cases (93.5%), and R1 margin in 2 cases (6.5%). Four cases (12.9%) were very low risk, 9 cases (29%) were low risk, 11 cases (35.5%) were intermediate risk, and 7 cases (22.6%) were high risk.

Of the 3 patients who underwent PHPD, the tumors were all located in the second portion, with a mean diameter of 3.3 (2.5–4.0) cm. None received neoadjuvant TKI, and 2 patients (66.7%) received postoperative adjuvant TKI. The mean operative time was 323 (300–350) min and the mean blood loss was 500 (400–600) ml. Biochemical leakage was observed in 1 case (33.3%) and no complications greater than Clavien-Dindo grade II were observed. The mean postoperative hospital stay was 12 (10–14) days. R0 margin was achieved in all the cases; 1 case (33.3%) was low risk while the other two (66.7%) were intermediate risk (Fig. 4). These three cases were summarized in Table 3.

Of the patients who received PD, the tumors were all located in the second portion, with a mean diameter of 5.2 (1.7–11.0) cm. One patient (2.9%) who received neoadjuvant TKI had an 11-cm-diameter tumor suspected of involving SMV, but the final surgical margin was also R1. Postoperative adjuvant TKI was performed in 11 cases (31.4%). The mean operative time was 354.9 (290–490) min and the mean blood loss was 582.9 (200–1300) ml. POPF was observed in 11 cases, of which 7 cases (20.0%) were biochemical leakage, 3 cases (8.6%) were grade B, and 1 case (2.9%) was grade C. DGE was observed in 11 cases, of which 2 cases (5.7%) were grade A, 5 cases (14.3%) were grade B, and 4 cases (11.4%) were grade C. Postoperative hemorrhage was observed in 3 cases (8.6%) which was cured by interventional therapy (Clavien-Dindo grade III). One patient (2.9%) died due to postoperative complications 125 days after surgery (Clavien-Dindo grade V). The mean postoperative hospital stay was 17.1 (5–125) days. R0 margin was achieved in 32 cases (91.4%), and R1 in 3 cases (8.6%).

Table 2 The demographic characteristics, tumor characteristics, and surgical outcomes

	Wedge resection	Segmental resection	PHPD	PD
Cases	23	31	3	35
Gender (male/female)	9/14	15/16	1/2	19/16
Age (years)	49.8 (13–71)	48.2 (26–74)	50.3 (38–65)	51.1 (34–71)
Site				
The first portion	9	10	0	0
The second portion	8	12	3	35
The third portion	6	4	0	0
The fourth portion	0	5	0	0
Tumor size (cm, the largest diameter)	2.3 (0.8–3.0)	4.5 (1.7–14.0)	3.3 (2.5–4.0)	5.2 (1.7–11.0)
Neoadjuvant TKI	0	1 (3.2%)	0	1 (2.9%)
Adjuvant TKI	4 (17.4%)	9 (29%)	2 (66.7%)	11 (31.4%)
Mean operative time (min)	212.6 (150–270)	260 (180–370)	323.3 (300–350)	354.9 (290–490)
Estimated blood loss (ml)	226.1 (100–400)	303.2 (100–600)	500 (400–600)	582.9 (200–1300)
POPF				
Biochemical leakage	1 (4.3%)	1 (3.2%)	1 (33.3%)	7 (20.0%)
Grade B	0	0	0	3 (8.6%)
Grade C	0	0	0	1 (2.9%)
DGE				
Grade A	2 (8.7%)	4 (12.9%)	0	2 (5.7%)
Grade B	1 (4.3%)	2 (6.5%)	0	5 (14.3%)
Grade C	0	2 (6.5%)	0	4 (11.4%)
Postoperative hemorrhage	0	1 (3.2%)	0	3 (8.6%)
Overall postoperative morbidity (Clavien-Dindo grade)				
I	3 (13.0%)	7 (22.6%)	2 (66.7%)	8 (22.9%)
II	1 (4.3%)	3 (9.7%)	0	6 (17.1%)
III	0	0	0	3 (8.6%)
IV	0	1 (3.2%)	0	0
V	0	0	0	1 (2.9%)
Postoperative hospital stay (day)	9.4 (5–25)	14.9 (5–102)	12 (10–14)	17.1 (5–125)
Surgical margin				
R0	21 (91.3%)	29 (93.5%)	3 (100%)	32 (91.4%)
R1	2 (8.7%)	2 (6.5%)	0	3 (8.6%)
Risk (NIH)				
Very low	3 (13.0%)	4 (12.9%)	0	3 (8.6%)
Low	8 (34.8%)	9 (29%)	1 (33.3%)	10 (28.6%)
Intermediate	6 (26.1%)	11 (35.5%)	2 (66.7%)	11 (31.4%)
High	6 (26.1%)	7 (22.6%)	0	11 (31.4%)

Three cases (8.6%) were very low risk, 10 cases (28.6%) were low risk, 11 cases (31.4%) were intermediate risk, and 11 cases (31.4%) were high risk.

In survival analysis, the wedge resection, segmental resection, and PHPD groups were merged into a limited resection group and compared with the PD group. At the end of the study, the median follow-up was 35 months, both the median RFS and OS had not been reached. The 1-, 3-, and 5-year RFS and OS of the entire cohort were 95.3%, 82.2%, 70.4%, and 97.6%, 89.8%, and 81.2%, respectively (Fig. 5A, C). Of the 57 patients in the limited resection group, 7 had relapsed (5 in the segmental resection group, 2 in the wedge resection group), 3 in liver, 3 in local, and 1 in both after a median recurrence-free interval of 30 months (range 4–48 months). Of the 35 patients in the PD group, 6 had relapsed, 3 in liver, 1 in local, 1 in local and liver, and 1 in local and peritoneum after a median recurrence-free interval of 22 months (range 4–42 months). The 1-, 3-, and 5-year RFS of the limited resection group and PD group were 96.0%, 82.8%, and 74.5% vs 94.0%, 81.7%, and 63.0%, respectively (log rank $p = 0.280$, HR 0.566, 95% CI 0.198–1.618, Fig. 5B). There

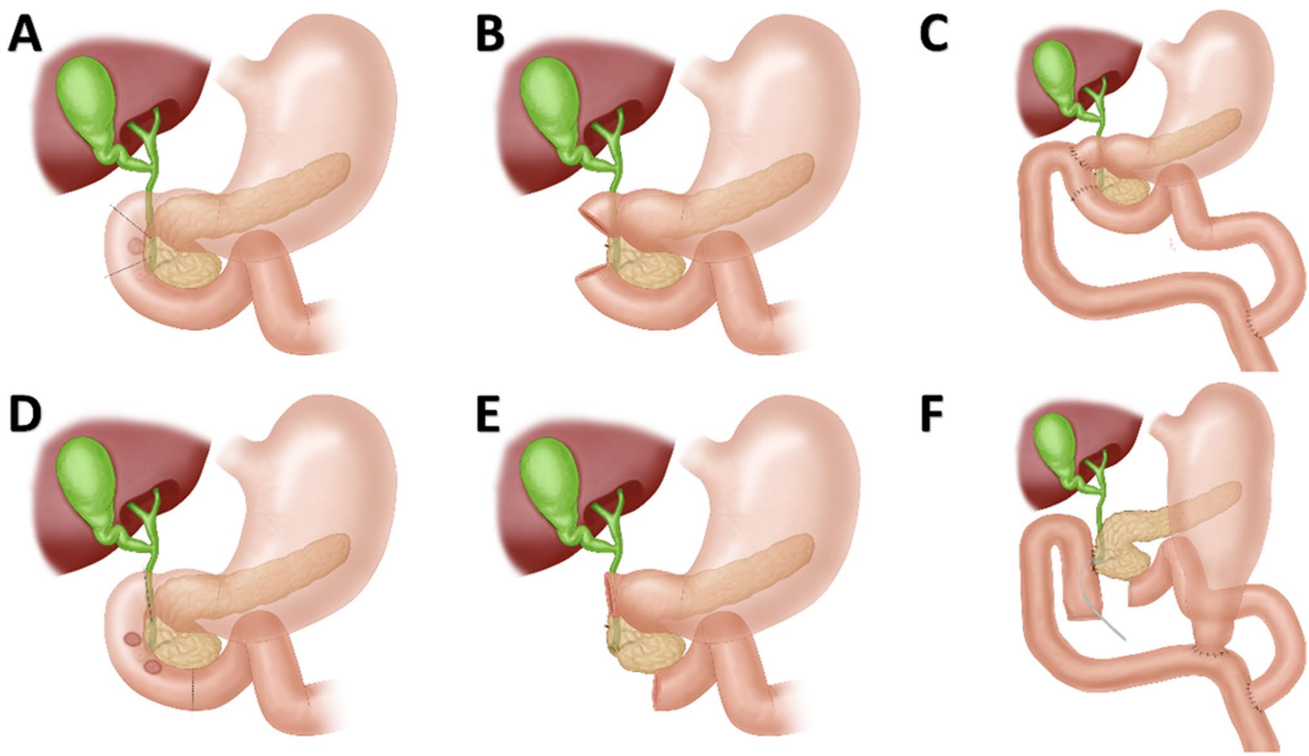

Fig. 4 **A**–**C** Segmental resection for the tumors in the proximal part of the second portion and reconstruction of digestive tract. **D**–**F** PHPD

Table 3 Results of the 3 cases of PHPD

	Case 1	Case 2	Case 3
Age	38	65	48
Tumor size (cm)	2.5	4.0	3.5
Operative time (min)	350	320	300
Estimated blood loss (ml)	500	600	400
POPF	-	Biochemical leakage	-
Overall postoperative morbidity (Clavien-Dindo grade)	-	I	I
Risk (NIH)	Low	Intermediate	Intermediate
Adjuvant TKI	-	+	+
Postoperative hospital stay	12	10	14
Recurrence	-	-	-
Follow-up (months)	42	35	20

were 4 deaths in the limited resection group (7%, 3 in segmental resection group, 1 in wedge resection group) and 4 deaths in the PD group (11.4%). The 1-, 3-, and 5-year OS of the limited resection group and PD group were 97.9%, 91.0%, and 83.4% vs 97.1%, 88.2%, and 77.2%, respectively (log rank $p = 0.469$, HR 0.602, 95% CI 0.150–2.412, Fig. 5D).

Discussion

GISTs are the most common mesenchymal tumors of the gastrointestinal tract, which can occur in any site of the gastrointestinal tract, but the stomach (60%) and small intestine (30%) are the most common primary sites. The duodenum (4–5%) and rectum (4%) are less common primary sites, while the esophagus (< 1%), colon, and appendix (1–2%) have only been reported in a few cases [3]. Liver metastases and/or intraperitoneal dissemination are the most common clinical manifestations of malignancy, and the incidence of nodal metastases is low. Distant metastasis is observed only in advanced cases [4].

Given the biological behavior characteristics of the GISTs, the principle goal of surgery is considered to aim at the removal of an intact tumor with a negative histological margin, and complex multiorgan resection should be avoided to minimize the surgical complications [4]. However, the complexity of the anatomy of the pancreaticoduodenal region makes the surgery for duodenal GISTs challenging.

PD is performed in many kinds of duodenal tumors; the head of the pancreas is removed even though the disease is located only in the duodenum, resulting in a high incidence of complications such as pancreatic fistula and hemorrhage [6]. Moreover, PD has a higher short-term morbidity for non-ampullary duodenal lesions, as they are usually associated with a soft pancreas and a thinner main pancreatic duct

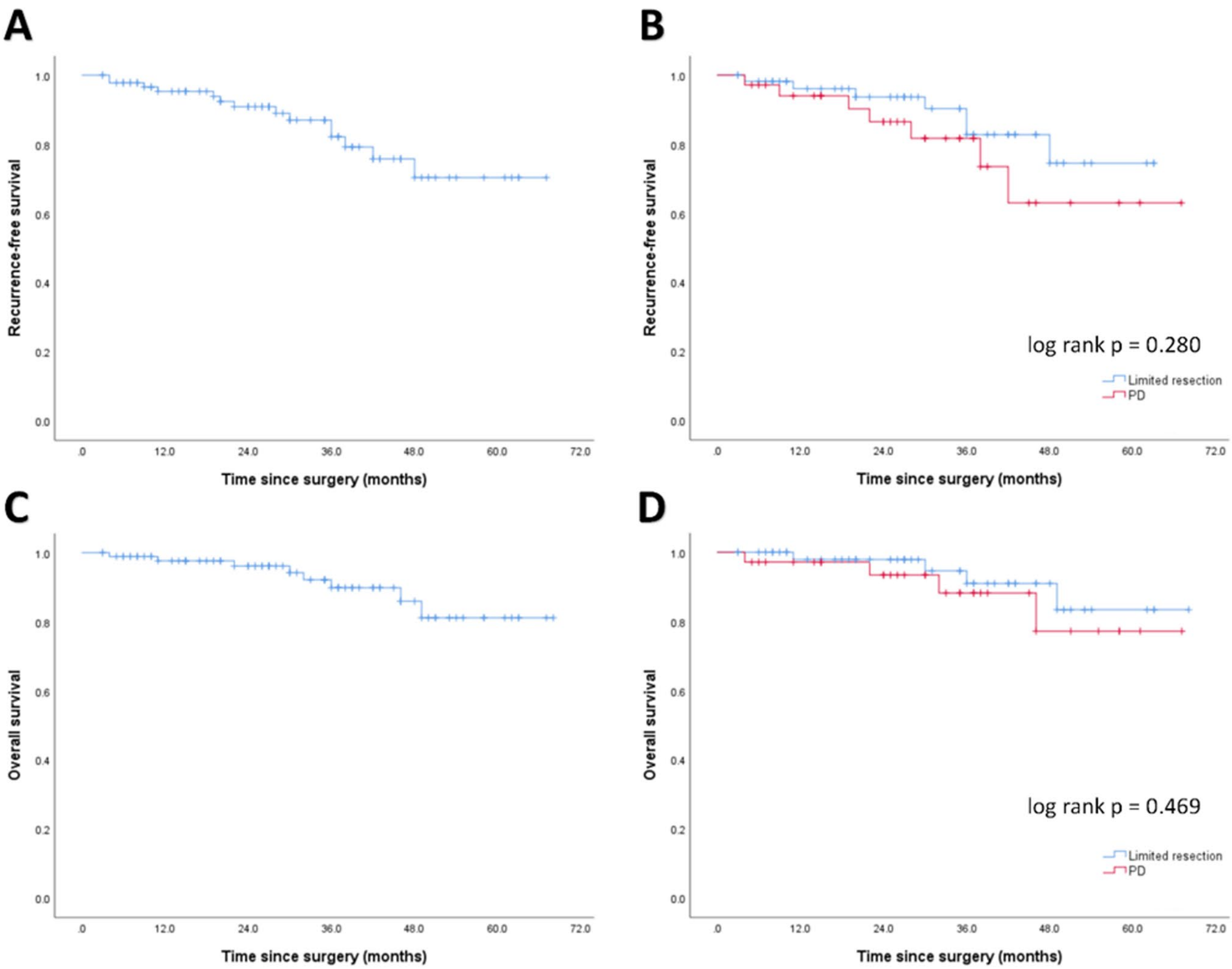

Fig. 5 **A**, **C** The RFS and OS of the entire cohort ($n=92$). **B**, **D** The RFS and OS of the limited resection group (blue, $n=57$) and PD group (red, $n=35$)

[12]. The long-term risks after PD are an increase of new onset diabetes and a significant decrease of exocrine functions [13].

Comprehensive treatment based on radical surgery for highly malignant tumors such as adenocarcinoma is the premise for improving long-term survival rate. For benign or low-grade malignant tumors, it is more important to reduce the scope of surgical resection, preserve important organs as much as possible and maintain the physiological continuity of the digestive tract to improve the quality of life after surgery. For duodenal GISTs, to achieve the goals of a histologically negative surgical margin, preserving organs and simplifying the surgical procedure as much as possible require a balance between PD and limited resection.

To summarize our experience of last 11 years, we believe that for the GISTs in all portions of the duodenum, if they are located away from the ampulla (> 1.5 cm) and without invasion of the pancreas, a wedge resection or a segmental resection is feasible. Many studies have pointed out that they preserve the pancreas; reduce operative time, postoperative morbidity, and mortality [14–17]; and allow for similar oncologic outcomes compared with PD [17–19]. But these studies have ignored an important issue, that is, a PD has to be performed for GISTs that invade the ampulla of Vater or the pancreatic head to obtain a negative margin.

However, we think PD may not be the optimal choice for the duodenal GISTs that are close to the ampulla (< 1.5 cm) but still without invasion of the ampulla of Vater and the pancreatic head. Pancreas-preserving total duodenectomy (PPTD) is a novel organ-preserving surgical procedure, which is first described in 1995 by Chung [20]. It is usually performed for benign or premalignant lesions, such as familial adenomatous polyposis, periampullary adenoma, and complex duodenal injury [20–23]. PPTD is a novel and complex surgical technique, through which the pancreas is completely preserved and long-term complications may be reduced.

Recent studies suggest that PPTD has similar short-term results to PD in terms of morbidity (such as pancreatic fistula, wound infection, delayed gastric emptying, and intraabdominal abscess) and mortality [21, 23]. But the reason for such results is probably that PD has been widely carried out for many years and the technique has been continuously improved, while PPTD is still not carried out much so far and the surgical technique is limited.

The other problem is that the entire duodenum is removed through PPTD even if the lesion is located only in the second portion. Therefore, we improved the PPTD to PHPD to remove only the first and second portions of the duodenum, thus reducing the surgical trauma and operation time. To our knowledge, PHPD performed for periampullary duodenal GISTs is rarely reported. For the 3 cases of the periampullary duodenal GISTs, we obtained intact tumors with negative margins and save the entire pancreas for the patient through PHPD. As surgical technique improves, PHPD may be performed more frequently for periampullary duodenal GISTs.

More recently, some researchers have proposed that enucleation is an option for elderly, frail patients with duodenal GISTs without mucosal ulcers, because of its lower medical risk and similar oncology outcomes [24]. However, there are few studies based on this aspect currently, and it is not included in the general recommendations. More researches are needed to verify it in the future.

Conclusions

The surgical strategies for duodenal GISTs should be based on tumor location, size, and invasion of the ampulla of Vater and pancreas. For duodenal GISTs without invasion of the ampulla of Vater or the pancreatic head, a limited resection (such as wedge resection, segmental resection or PHPD) preserves the pancreas which may reduce early and late postoperative complications compared with PD. But for duodenal GISTs with an invasion of the ampulla of Vater or the pancreatic head, a PD is still necessary in order to achieve a negative margin. For duodenal GISTs in different locations, we should strike a balance between PD and limited resection to maximize patient benefit.

Acknowledgements We would like to thank the National Comprehensive Cancer Network (https://www.nccn.org/) for the NCCN guideline of gastrointestinal stromal tumors. We would like to thank 1·3·5 Project for Disciplines of Excellence Clinical Research Incubation Project for the financial support.

Authors' contributions Xiang Fu: study conception and design, acquisition of data, analysis and interpretation of data, drafting of manuscript. Xing Wang: study conception and design, acquisition of data, analysis and interpretation of data, drafting of manuscript. Junjie Xiong: acquisition of data and analysis and interpretation of data. Yutong Yao: acquisition of data and analysis and interpretation of data. Chunlu Tan: acquisition of data, analysis and interpretation of data, critical revision of manuscript. Xubao Liu: acquisition of data, analysis and interpretation of data, critical revision of manuscript.

Funding This study was supported by the 1·3·5 Project for Disciplines of Excellence Clinical Research Incubation Project (ZY2017302), West China Hospital, Sichuan University.

Availability of data and material The data used or analyzed during the current study are available from the corresponding author on reasonable request.

Code availability Not applicable.

Declarations

Ethics approval This study was approved by the local ethical review board of the West China Hospital.

Consent to participate Informed consent for participation was obtained from the patients who were involved in the study.

Consent for publication Written informed consent for publication was obtained from all participants.

Conflict of interest The authors declare no competing interests.

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