Brief Report

Duodenoduodenostomy in the Treatment of Superior Mesenteric Artery Syndrome

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Superior mesenteric artery syndrome (SMAS) is a rare and morbid disease affecting .1 to .3% of the population.[1] Its pathology is due to a mechanical obstruction at the level of the 3rd portion of the duodenum as the SMA pedicle traverses over it before expanding into redundant folds of mesentery. Current treatment standards consist of weight gain to build up the mesenteric fat pad to increase the distance of the aorto-mesenteric angle. However, when this fails, surgery is the next option.

Current operative techniques are primarily focused on either bypassing the obstruction or releasing the duodenum from compression. Unfortunately, these procedures have a high failure rate partially due to their unique technical and anatomic constraints. The goal of any new operation should relieve the obstruction of the duodenum, preserve the duodenum's functional anatomy, and prevent re-entrapment. This report describes a novel operation for SMAS which fulfills these criteria and the second patient who has undergone this procedure. The operation is the medial rotation of the duodenum with duodenoduodenostomy.[2]

The patient is a 40-year-old woman who began having digestive symptoms of intermittent obstruction at the age of 18 years. Her pre-SMAS past medical history was negative for endocrine, autoimmune, neoplastic, and psychiatric disorders. At the age of 34 years, she progressed to near obstruction and was unable to tolerate solid foods. As a result, she underwent multiple procedures including gastrostomy tube placement for decompression and comfort, a separate jejunostomy tube placement for enteral nutrition, and subclavian port placement for parenteral nutrition. She was referred to our clinic through a support network for people with SMAS. By the time she came for her clinic visit, she had registered for hospice for chronic pain and failure to thrive. Her presenting weight was 82 lbs with her normal pre-SMAS weight at 115-120 lbs.

The operation was performed through a midline laparotomy carried out from the epigastric region to just inferior to the umbilicus. The retroperitoneum of the right colon and small intestine was divided. Starting with the 4th portion of the duodenum, the duodenum was then mobilized toward the head of the pancreas past the SMA/superior mesenteric vein (SMV) pedicle. A key step here is to mobilize the duodenum all the way to the first branch of pancreatico-duodenal arcade. This will bring the 3rd portion of the duodenum just to the right of the SMA pedicle, and the SMV is seen immediately to the left (Figure 1). At this point, the small bowel goes from being midline to falling away toward the patient's right. As the 4th portion of the duodenum and proximal jejunum is brought to the right side of the patient, there is a slight clockwise rotation of the mesentery. The duodenoduodenostomy is performed and this anchors the mesentery in place to prevent further rotation, and small bowel is comfortably repositioned to the patient's right side (Figure 1). The repositioning of the 3rd and 4th portions of the duodenum maintains normal anatomic function and prevents either the duodenum or jejunum from being entrapped by the SMA pedicle. A nasojejunal feeding tube was placed for early enteral nutrition and to allow the anastomosis to heal before stressing it with a *per os* challenge (Figure 2C). On POD 5, an upper gastrointestinal study with small bowel follow-up showed a patent anastomosis with normal transit. On the same day, she was started on liquids and her nasojejunal tube was removed. On POD 8, she was given a soft mechanical diet. This was the first solid meal she had in 5 years. She has since progressed to a regular diet without pain from eating. At 8 weeks follow-up, her weight increased from 82 lbs to 93 lbs without supplementation and enjoying all foods.

In 2017, we performed the first duodenoduodenostomy operation to treat SMAS for a patient with near obstruction who was also not able to tolerate solid foods and had a 50 lb weight loss.[1] Nearly 3 years after surgery, the patient has now a normal Body Mass Index of 20.9, and her last pre-albumin measured 2 years after surgery was 39 mg/dL. This patient has also not had recurrence of SMAS or required admission for primary bowel-related issues. Our second patient presented in this report had a similar clinical presentation, except her preoperative

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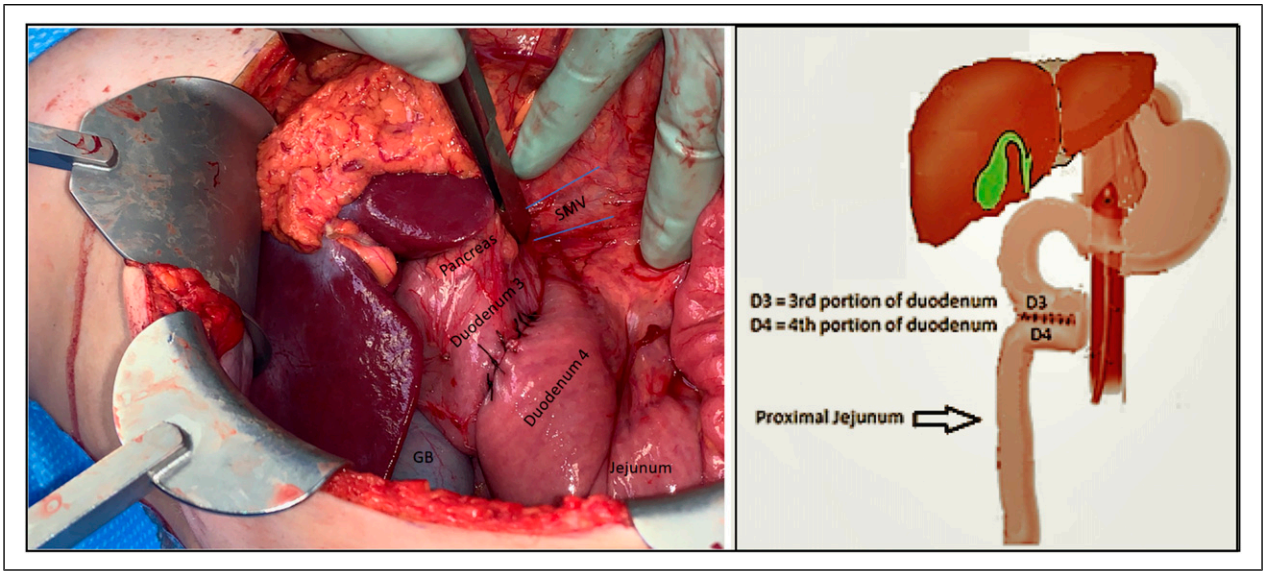

Figure 1. Operative photo of the duodenoduodenostomy and illustrative diagram of relational anatomy. GB, gallbladder; SMV, superior mesenteric vein.

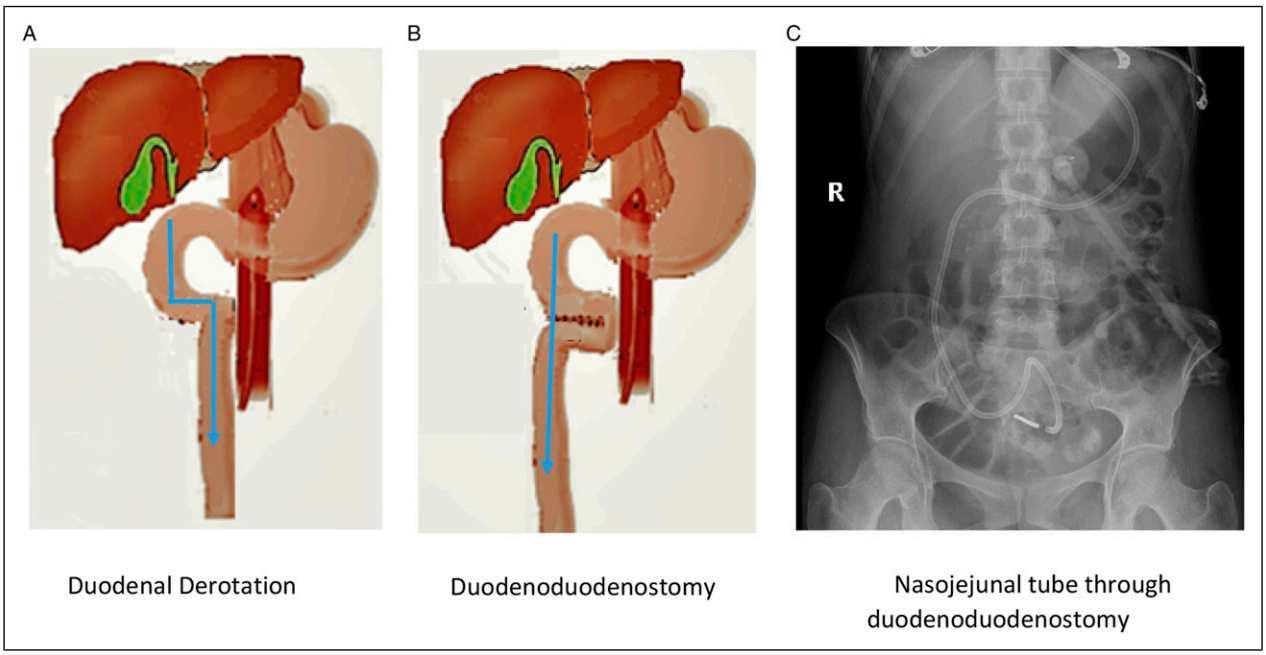

Figure 2. Transit through the duodenum. (A). Duodenal derotation - Pathway through duodenal derotation procedure. (B). Duodenoduodenostomy - Pathway through duodenoduodenostomy. (C). Nasojejunal tube through duodenoduodenostomy - Patient's abdominal X-ray with nasojejunostomy feeding tube through duodenoduodenostomy.

condition appeared worse because she had transitioned to hospice for failure to thrive. Her operation has given us a better understanding of the procedure by validating the clinical results of our first operation. It has also provided an opportunity to better articulate the key operative steps and the rationale behind them.

It is important to acknowledge that the diagnosis and treatment of SMAS is challenging. SMAS is rare, and it can be confused with other foregut pathology.[3] It also carries a high incidence of psychiatric comorbidities, which may add to an extensive workup and ultimately a delay in care. However, when inadequately diagnosed or

ignored, SMAS can be life-threatening. Current operative therapies have evolved from relieving the obstruction by de-rotation procedures to now bypassing the obstruction all together. Unfortunately, both types of procedures have their own unique limitations based on the nature at which they address the obstruction at the 3rd portion of the duodenum.

The duodenal mobilization techniques such as Strong's procedure and duodenal de-rotation have a risk of recurrence likely because of 3 things. First, a significant portion of the 3rd portion of the duodenum is tethered to the head of the pancreas, due to sharing the same blood supply, and may not clear the SMA pedicle. This can be seen in Figure 1, where the apex of the duodenoduodenostomy is just medial to the SMV. Second, the C-loop of the duodenum results in a natural sweep pointing to the patient's left. Because of this, the duodenum may migrate beneath the SMA pedicle. Third, by removing the suspensory ligament of Treitz, the small bowel will create a sharp turn at the point where the 3rd portion of the duodenum is tethered by the pancreas (Figure 2A). The duodenoduodenostomy, on the other hand, changes the direction of the sweep of the small bowel. The anastomosis is well to the right of the patient and away from the SMA pedicle. The duodenoduodenostomy also creates a straight path down from the proximal duodenum to jejunum, allowing for a lower path of resistance for the transit of food (Figure 2B). This path can be appreciated in an X-ray showing a nasal jejunal feeding tube pointing straight down the patient's right side without turning past midline (Figure 2C). As for bypass procedures, the preferred operation for SMAS is the duodenojejunostomy. However, in the only study to examine post-discharge follow-up for SMAS after duodenojejunostomy, they reported a failure rate of 67%.[4] Possible causes for failure may be due to the fact that the obstruction is not relieved and may be a constant source of pain. In addition, the bypassed proximal jejunum can act as a p-trap, similar to the plumbing of a sink, predisposing patients to potential bacterial overgrowth.

In summary, the duodenoduodenostomy is a novel operation for the definitive treatment for SMAS. It is unique in that it relieves the obstruction of the duodenum, while maintaining its anatomic function and repositions the distal duodenum and proximal small bowel to prevent re-entrapment. The preliminary findings are promising for providing immediate relief of SMAS-related obstruction with return of a normal and regular diet. As with other operative procedures for SMAS, the duodenoduodenostomy should be reserved for patients who have failed medical management. Future studies will be needed to examine longer term outcomes.

Declaration of Conflicting Interests

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