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Surgical treatment of congenital hyperinsulinism

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Abstract

Objectives: This study aims to analyze the details of surgical techniques performed in various types of congenital hyperinsulinism (CHI) and review our experience.

Patients and methods: Six children with CHI (3 males, 3 females) treated between January 2013 and January 2024 were retrospectively reviewed.

Results: Four patients had diffuse type CHI, and two had focal type CHI. The median age at diagnosis was 18.5 days (range, 1 to 54 days). The diagnosis involved 18F-DOPA (18-fluoro-dihydroxyphenylalanine) positron emission tomography (PET)/computed tomography (CT) in two patients and 68Ga-NODAGA-exendin-4 PET/CT in one patient. Mutations in ABCC8/KCNJ1 were detected in five patients. Ultrasonography (n=2), frozen biopsy (n=2), and indocyanine green-guided identification (n=1) were used intraoperatively. Near total pancreatectomy (95 to 98%) and gastrostomy were performed in four patients with diffuse CHI. Among the two patients with focal lesions, distal pancreatectomy was performed in one patient, and enucleation was performed in the other patient.

Conclusion: Knowing the technical details of the surgery, preparation for different surgical approaches to various types of CHI, and being aware of the risky major complications achieve successful outcomes in pancreatic surgery.

Keywords: Children, congenital hyperinsulinism, pancreatectomy.

Congenital hyperinsulinism (CHI) is a rare condition occurring in 1 in 20,000 live births.[1] The disease is characterized by inappropriate insulin secretion from pancreatic β-cells, which results in a decrease of plasma glucose concentrations during fasting or failure to increase insulin secretion as a response to glucose loading.[2] Congenital hyperinsulinism is a neonatal clinical emergency because rapid control of recurrent hypoglycemia is important to prevent long-term irreversible severe neurological damage in infants.[1] Patients with CHI may present with diffuse involvement of the pancreatic β-cells (diffuse hyperinsulinism) or focal adenomatous islet cell hyperplasia (focal hyperinsulinism).[3] Previously the disease was described as "nesidioblastosis," as it was considered a developmental abnormality, resulting excess insulin secretion from high numbers of pancreatic islet cells.[4] However, molecular studies have shown that genetic derangements affecting insulin/glucose homeostasis are the main pathology.[5] The most severe form of CHI is caused by inactivating mutations of ABCC8 and KCNJ11 genes encoding the subunits of the ATP-sensitive potassium channels (K_{ATP}), which are expressed in pancreatic β-cells.[2] While biallelic recessive or dominant mutations cause diffuse CHI, inheritance of a paternal recessive mutation together with a focal loss of heterozygosity causes focal CHI.[4]

Congenital hyperinsulinism is diagnosed based on the following criteria: fasting hypoglycemia associated with inadequate supression of plasma insulin level and evidence of low plasma beta-hydroxybutyrate and free fatty acids, accompanied by an inappropriate glycemic response to glucagon stimulation.[4] The surgical decision is made according to the patient's response to medical treatment. If medical treatment fails, surgical options should be considered according to the type

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of CHI (diffuse/focal) based on genetic tests and imaging studies.[4,6]

The main goal of surgical treatment is to preserve normal pancreatic tissue as much as possible for endocrine and exocrine functions and remove the diseased pancreatic tissue as safely as possible. While near-total pancreatectomy (95 to 98%) is preferred in patients with diffuse disease, local resection or enucleation of the involved portion of pancreas could be curable operations for focal lesions with low risk of diabetes mellitus.[7,8]

Due to the complexity and multivariate nature of surgical treatment in CHI, these patients are followed up only in centers with requisite expertise. Therefore, data regarding optimal work-up and surgical management strategies on this topic is limited in the literature. Hence, this study aimed to analyze the details of surgical techniques performed in various types of CHI and review our experience.

PATIENTS AND METHODS

Data of patients with CHI treated at the Marmara University Faculty of Medicine between January 2013 and January 2024 were retrospectively reviewed. Over the 10-year study period, eight patients with CHI were identified. Since two patients were operated in different centers, they were excluded from the study. Six patients (3 males, 3 females) were included in the final analyses.

The parameters used in diagnosis were fasting hypoglycemia (glucose <50 mg/dL), plasma insulin >2 μU/mL, low plasma beta-hydroxybutyrate (<1.8 mmol/L), low free fatty acids (<1.7 mmol/L), and inappropriate glycemic response to intravenous glucagon administration (≥30 mg/dL increase). Medical treatment was started after the disease was diagnosed according to previously described criteria. Medical treatment included continuous glucose infusion, frequent oral feeding, and the use of diazoxide, glucagon, or octreotide to stabilize blood glucose level. Careful preoperative planning was conducted due to the unique physiological and metabolic challenges in CHI. Drug doses of medical treatment and detailed preoperative, intraoperative, and postoperative considerations are presented in Table 1.

Surgical treatment was required in patients who did not respond to medical treatment and had severe hypoglycemia. Surgical approach was chosen according to the type of CHI (diffuse/focal). Patients were screened for genetic mutations of ABCC8 and KCNJ11, which encode the subunits of the KATP channel. In patients thought to have a focal lesion according to genetic analysis, 18-fluoro-dihydroxyphenylalanine (^{18}F-DOPA) positron emission tomography (PET)/computed tomography (CT) and ^{68}Ga-NODAGA-exendin-4 PET/CT were performed as imaging studies.

A written informed consent was obtained from the parents and/or legal guardians of the patients.

TABLE 1
Medical treatment and phases of surgical preparation

Preoperative considerations	Associated conditions	Medical treatment drug/dose	Intraoperative considerations	Postoperative considerations
Correct electrolyte imbalances	Cardiomyopathy	**Diazoxide** 2-5 mg/kg/day (initial) 2-3 divided doses 20 mg/kg/day (maximum)	Continuous blood glucose monitoring (every 15-30 min)	Monitor glucose frequently in ICU (every 1-2 h initially)
Stabilize blood glucose levels	Neurological damage	**Octreotide** 5-30 μg/kg/day 6-8 hourly, subcutaneous injection	Dextrose containing fluids Adjust based on intraoperative glucose monitoring Electrolyte replacement	Hyperglycemia risk (following partial/total pancreatectomy due to reduced insulin secretion)
Family counseling	Nutritional status	**Glucagon** 5-20 μg/kg/h 2.5-5 μg/kg/h (starting dose)	Be prepared for significant blood loss Manage heat loss, prevent hypothermia	Hypoglycemia risk (persistent hypoglycemia if surgery does not completely resolve the condition)

ICU: Intensive care unit.

The study protocol was approved by the Marmara University Faculty of Medicine Ethics Committee (date: 20.09.2024, no: 09.2024.964). The study was conducted in accordance with the principles of the Declaration of Helsinki.

Surgical technique

The surgical technique was determined based on preoperative diagnostic studies, as well as perioperative findings. Surgical strategies varied depending on whether CHI was diffuse or focal. All operations were performed via a transverse supraumblical laparotomy. In patients with likely diffuse CHI or with no focal lesions localized preoperatively, near-total pancreatectomy (95 to 98%) was the preferred surgical approach, which involved the resection of the tail, body, uncinate process, and part of pancreatic head, leaving only a rim of pancreatic tissue between the common bile duct (CBD) and the duodenal wall. First, the pancreas was completely exposed with an extended Kocher maneuver, and the inferior border of the pancreas was mobilized by entering the lesser sac through the gastrocolic omentum. The tail of the pancreas was then carefully dissected away from the hilum of the spleen, and the splenic artery and vein were identified and preserved (Figure 1). During the dissection, in a medial direction towards the head of the pancreas, short pancreatic vessels passing from the splenic vessels into the pancreas were divided using bipolar diathermy. After arriving at the superior mesenteric vessels, a sling was placed around them to retract the vessels to ensure adequate mobilization of the uncinate process. To perform a true near-total pancreatectomy, CBD was skeletonized with meticulous care, and a vessel loop was placed around the extrapancreatic section of the CBD posterior to the duodenum (Figure 2). During subsequent dissection the pancreatic head, swinging the vessel loop within the C-loop of the duodenum guided the position of the CBD. The head of the pancreas was mobilized, and superior and inferior pancreaticoduodenal vessels were divided. Pancreatic duct was ligated with nonabsorbable sutures and divided with LigaSure™ (Medtronic, Minneapolis, MN, USA). At the end of the dissection, the gallbladder was milked to identify any bile leak to ensure that there was no CBD injury. A gastrostomy tube was also placed for possible need for enteral access in babies with diffuse CHI.

In patients with a focal lesion preoperatively, that portion of the pancreas was initially exposed. Focal lesions are difficult to distinguish from normal pancreatic tissue;[3] therefore, the pancreas was inspected under loupe magnification and palpated carefully to identify the focal lesion (usually 10 mm or less in diameter). Small and superficial lesions in the body or tail were treated by distal pancreatectomy or enucleation. Similarly, if there was a superficial focal lesion located in the head of the pancreas, enucleation was the preferred technique. When focal lesions are buried within the pancreas, they are impossible to observe and palpate.[3] Therefore, intraoperative ultrasonography (US) and indocyanine green (ICG)-guided identification of the accurate location

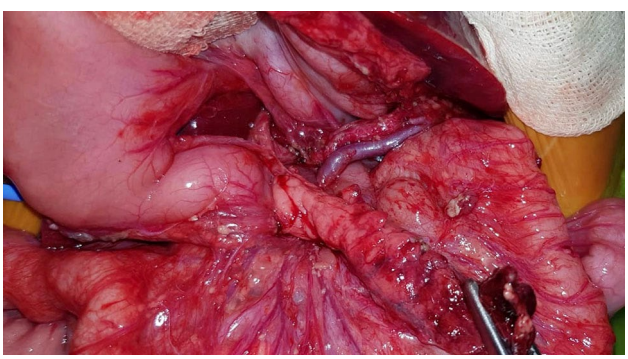

Figure 1. Operative view of the skeletonized splenic artery and vein, after dissection of the tail of the pancreas.

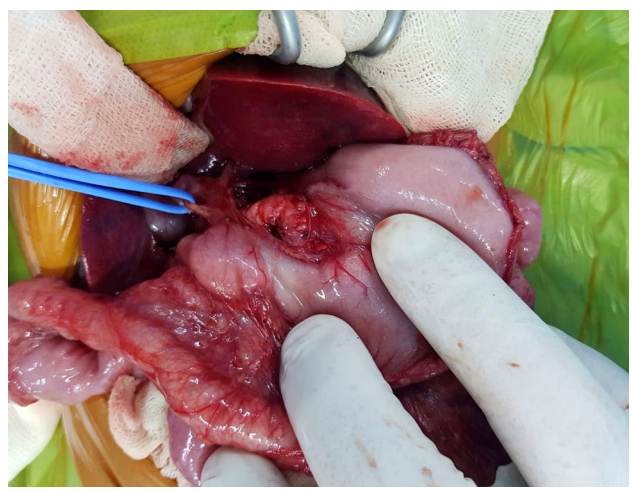

Figure 2. Operative view of the vessel loop placed around the extrapancreatic section of the CBD posterior to the duodenum.

CBD: Common bile duct.

of focal lesions were additional imaging methods used during the operation. Intraoperative frozen section samples were also sent for clear margins after local resection of focal lesions. If deep focal lesions were located in the body and tail of the pancreas, distal pancreatectomy was required for complete resection. In more severe cases, if there was a deep focal lesion in the pancreatic head or extending into the duodenal wall, after complete resection of the pancreatic head, drainage of the remaining body or tail with Roux-en-Y pancreaticojejunostomy or Whipple procedure was performed. A drain was placed at the end of all operations performed in various types of CHI.

Statistical analysis

This was a descriptive study reflecting our experience on six patients with CHI. Continuous variables are presented as median and range. Categorical variables are presented as n and %.

RESULTS

Four patients had diffuse type CHI, and two had focal type CHI. All patients were operated by a single surgeon, and five of them had undergone surgery within the last four years. Preoperative genetic tests, imaging studies, and final pathology were used to establish a diagnosis. Median age at diagnosis was 18.5 days (range, 1 to 54 days). Mutations in ABCC8/KCNJ1 were detected in five patients with genetic analysis. Two patients underwent ^{18}F-DOPA PET/CT for accurate localization of the focal lesion (Figure 3). One patient had a focal lesion in the body/tail of the pancreas, and the other one had a focal lesion in the head of the pancreas. The exact location of the focal lesion in the head of the pancreas was also confirmed with ^{68}Ga-NODAGA-exendin-4 PET/CT in one patient. Intraoperative USG was also used in these patients; however, only the focal lesion located in the body/tail of the pancreas could be determined precisely (Figure 4). In addition, intraoperative frozen biopsies were taken in two patients with focal lesions; however, no definitive results were obtained in the pathological examination. Moreover, ICG was also used in one patient who had a focal lesion in the head of the pancreas; however, we were unable to determine the exact location of the lesion, most likely due to technical problems.

Near total pancreatectomy (95 to 98%) and gastrostomy were performed in all four patients with diffuse CHI. According to the outcomes, persistent hypoglycemia was controlled with enteral nutrition, and reduced octreotide doses were required during follow-up. Furthermore, life-threatening hypoglycemia attacks were prevented in these patients who were closely monitored. Nevertheless, one of these patients presented with persistent vomiting two months

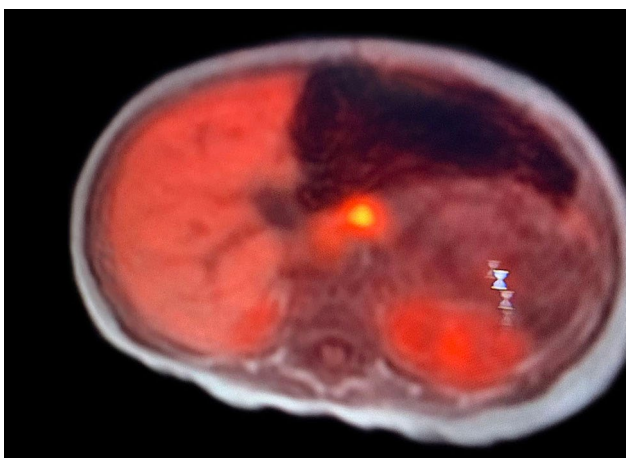

Figure 3. Focal lesion located in the body-tail of the pancreas with ^{18}F-DOPA PET/CT.

^{18}F-DOPA: 18F-Fluoro-dihydroxyphenylalanine; PET/CT: Positron emission computed tomography.

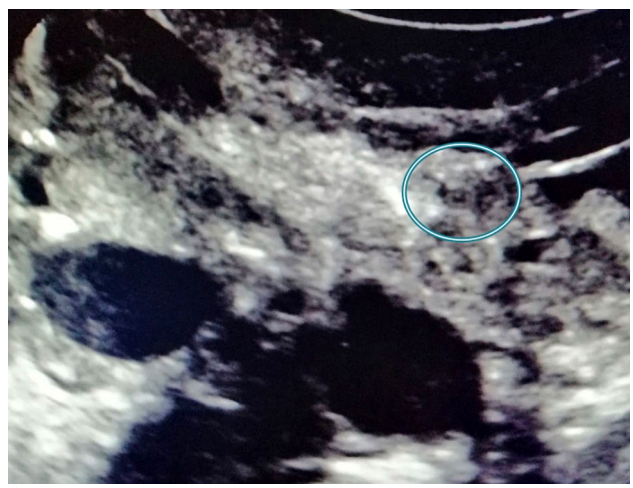

Figure 4. Intraoperative USG of the focal lesion circled in the body/tail of pancreas.

USG: Ultrasonography.

TABLE 2
Patient characteristics, diagnostic studies, surgical procedure, complications, and outcome

Patient	Age at diagnosis (Day)	Age/Sex	Pathology	Preoperative diagnostic studies Imaging studies genetic tests	Intraoperative diagnostic studies	Surgical procedure (Age at the time of surgery)	Complications (Postoperative time)	Outcomes/medication
1	30	Female	DCHI	Homozygous KCNJ11 mutation	-	Near total pancreatectomy+gastrostomy (6 months)	-	Octreotide 95 mg/month, creon, avicap, evicap, polivit
2	7	Male	DCHI	Homozygous ABCC8 mutation	-	Near total pancreatectomy+gastrostomy (2 months)	-	Octreotide 20 mg/month, creon, avicap, evicap, polivit, clexane
3	31	Male	FCHI	^{18}F-DOPA PET/CT ^{68}Ga-NODAGA-exendin-4 PET/CT Heterozygous ABCC8 mutation	Frozen biopsy US ICG	Enucleation (7 months)	-	Completely cured No medication
4	54	Male	DCHI	Homozygous KCNJ11 mutation	-	Near total pancreatectomy+gastrostomy (3 months)	-	Octreotide 20 mg/month creon, avicap, evicap, polivit
5	1	Female	FCHI	^{18}F-DOPA PET/CT Heterozygous ABCC8 mutation	Frozen biopsy US	Distal pancreatectomy+gastrostomy (4 months)	Persistent vomiting (4 months) (follow-up in PICU)	Octreotide 20 mg/month, creon, avicap, evicap, polivit Motilium, gaviscon, kepra **Exitus** (Severe hypoglycemia due to vomiting)
6	1	Female	DCHI	Data not available (first patient of the study)	-	Near total pancreatectomy+gastrostomy (2 months)	Persistent vomiting, ileus Second operation for brid ileus (2 months) (follow-up in PICU)	Octreotide 20 mg/month creon, avicap, evicap, polivit **Exitus** (Postoperative sepsis)

DCHI: Diffuse congenital hyperinsulinism; FCHI: Focal congenital hyperinsulinism; ^{18}F-DOPA: ^{18}F-Fluoro-dihydroxyphenylalanine; PET/CT: Positron emission computed tomography; US: Ultrasonography; ICG: Indocyanine green; PICU: Pediatric intensive care unit.

after discharge. Patient underwent surgery for brid ileus and died due to sepsis postoperatively.

Regarding the surgical techniques performed in two patients with focal CHI, distal pancreatectomy and enucleation were the choice of treatment. In one patient with a focal lesion in the body/tail of the pancreas, the lesion could not be visualized or palpated. Intraoperative US was used in this patient, and the deep location of the focal lesion in the body/tail of the pancreatic tissue was detected. Distal pancreatectomy was performed in this patient. In the other patient who had a focal lesion in the head of the pancreas, a 11-mm focal lesion, which had a marble-like appearance, was identified under loupe magnification. The superficial focal lesion was also firmer in texture with palpation, and enucleation was performed using tenotomy scissors rather than cautery. Analyzing the surgical outcomes of focal lesions, one patient was completely cured. Enucleation of the focal lesion in the head of the pancreas was the surgical technique performed in this patient. The patient did not require postoperative medical treatment. Moreover, glucose levels were normalized as soon as the focal lesion was removed during surgery. In the other patient, who had distal pancreatectomy for focal lesion located in the body-tail of the pancreas, the doses of medication were significantly reduced postoperatively. However, the patient died due to persistent vomiting and secondary hypoglycemia four months after discharge.

Age at diagnosis, sex, diagnostic studies, surgical procedure, age at the time of surgery, complications, postoperative time, and outcomes are presented in Table 2.

DISCUSSION

Congenital hyperinsulinism presents diagnostic and therapeutic challenges. The decision for surgery is based on the patient's response to medical treatment, imaging results, and genetic testing. Since noninvasive conventional imaging methods (ultrasonography, computed tomography, and magnetic resonance) or invasive methods (arterial stimulation with venous sampling and transhepatic portal venous sampling) have not been successful in diagnosis, ^{18}F-DOPA PET/CT scan is accepted as the current standard imaging method for distinguishing focal and diffuse CHI.[4] Radiotracer uptake is visually observed to be higher in the area thought to be a focal lesion than in other parts of the pancreatic tissue. If uniform uptake is observed throughout the pancreatic tissue, it is considered diffuse disease.[4] It has also been observed that it is useful in demonstrating possible ectopic lesions in a location other than the pancreas.[9] In some experienced centers, ^{18}F-DOPA PET/CT is performed in each patient regardless of the genetic analysis result, while in algorithms presented in the latest published studies ^{18}F-DOPA PET/CT scan is performed only in patients thought to have a focal lesion according to genetic analyses.[3,6] On the other hand, ^{18}F-DOPA PET/CT scan had only 85 to 89% sensitivity in detecting focal CHI.[10] Since focal lesions were missed in some patients with ^{18}F-DOPA PET/CT, another imaging method has been decribed recently.[10,11] Authors reported that ^{68}Ga-NODAGA-exendin-4 PET/CT has a higher clinical sensitivity and better interobserver correlation than ^{18}F-DOPA PET/CT.[11] Therefore, in our study, although we determined the location of the focal lesion with ^{18}F-DOPA PET/CT in two patients, we additionaly performed ^{68}Ga-NODAGA-exendin-4 PET/CT in one patient. The exact location of the focal lesion in the head of the pancreas was also confirmed with ^{68}Ga-NODAGA-exendin-4 PET/CT; hence, we preoperatively improved our surgical strategies for this difficult location, which could complicate the surgery.

While near-total pancreatectomy applied in diffuse CHI does not provide a complete cure in these patients, it can help prevent severe hypoglycemia and brain damage.[4] Even in experienced centralized centers, up to 50% of patients continue to have hypoglycemia or develop diabetes mellitus and exocrine insufficiency during follow-up.[3,12] As emphasized in multiple studies, deciding on the extent of pancreatic resection to be performed and the pancreatic tissue to be preserved are decided by intraoperative pathological examination.[13] While some authors prefer the resection of the tail of the pancreas for urgent frozen section analysis to confirm the diagnosis,[1] another experienced center preferred multiple 2- to 3-mm biopsies taken sharply with tenotomy scissors from the pancreatic head, body, and tail.[4] However, in both surgical approaches, further resection only commences after the

diagnosis of diffuse CHI is confirmed.[1,4] Further pancreatic resection is dependent on the results of frozen samples according to current studies; however, in our study, surgical decisions were determined according to preoperative clinical course of patients with diffuse CHI due to lack of experience in pathological examination.

Since high risk of diabetes and neurobehavioral deficits were reported in patients who underwent near-total pancreatectomy, localized resection of a focal lesion is recommended if a focal CHI is likely.[14] Furthermore, histologic evidence of a focal lesion was actually observed more than expected when the pancreatectomy specimens were examined retrospectively.[3] Unfortunately, focal lesions are difficult to distinguish according to appearance and structure compared to normal pancreatic tissue. They may present slightly reddish or have a marble-like appearance in color and are often palpated slightly firmer in texture.[4] In addition, if a focal lesion is not visualized, taking samples from suspicious areas for frozen section analysis is essential until a focal lesion is found.[4] Partial pancreatectomy can be performed once the focal lesion is identified. However, before concluding the surgery, it is necessary to confirm the clear surgical margins with intraoperative frozen section due to the irregular shape of the lesions.[3] Likewise, in our study intraoperative frozen section samples were sent for clear margins after local resection of focal lesions. However, operations were concluded according to the surgeon's decision since the results of pathological examination were inaccurate. This controversial issue was deliberately highlighted in this study, to draw attention to a common problem in Türkiye. According to the literature, these surgeries have been progressing with frozen section results for over a decade.[13] Although our hospital is an advanced center and has a pathologist who is specialized in the pancreas, they hesitate to provide frozen section results during these surgeries. According to their analysis, the inadequate results of frozen section analysis are caused by two debatable issues. Although our institution is a reference center for these complicated surgeries, they believe that they have insufficient experience for accurate decision with frozen section analysis during surgery. Since there is no centralization in rare diseases in Türkiye, the number of cases encountered is still low for advanced experience. The second reason is the limitation of performing immunohistochemical study for frozen section analysis, which encourages pathologists for accurate decisions.

When focal lesions are buried within the pancreas, it is impossible to observe or palpate them. If the lesion has a pseudocapsule, it may sometimes be possible to detect it with intraoperative US. Since most focal lesions have similar echogenicity to normal pancreatic tissue, localization of the lesion is often unsuccessful. However, use of intraoperative US to delineate the pancreatic duct is still recommended, which is only 0.3 to 0.4 mm in diameter, as possible injury will be prevented by identifying the course of the tiny pancreatic duct.[3] In our study, we determined the exact location of the focal lesion with intraoperative US only in one patient. Since we initially used this additional imaging method only in patients with focal lesions, we now believe that intraoperative ultrasound should be employed for all patients with various types of CHI to accurately delineate the pancreatic duct during surgery.

Another challenging situation in pancreatic surgery is resection of deep focal lesions located at the head of the pancreas. Laje et al.[15] reviewed their outcomes in these complicated cases who underwent complete resection of the pancreatic head and Roux-en-Y pancreaticojejunostomy to the remaining pancreatic body/tail that is not amenable to local resection. In their study, the authors conclude that drainage of the remaining pancreatic body/tail with Roux-en-Y pancreaticojejunostomy is a safe and effective procedure.[15] Since the main goal is to preserve normal pancreatic tissue as much as possible, we were prepared for a possible Roux-en-Y pancreaticojejunostomy in the patient who had a focal lesion in the head of the pancreas. During exploration of the pancreatic tissue, we found a superficial focal lesion in the head of the pancreas, and it was removed with enucleation successfully.

There are two published reports on laparoscopic pancreatectomy for CHI.[16,17] The decision regarding laparoscopic versus open approach is at the discretion of the operating surgeon. Given the technical challenge of preserving the duodenum and CBD during pancreatic head

resection, in patients with diffuse CHI, near-total pancreatectomy (98%) is best approached via open laparotomy. Moreover, Adzick et al.[3] emphasized in their study that they had to reoperate babies with diffuse CHI who underwent inadequate laparoscopic pancreatectomy performed at other hospitals. On the other hand, laparoscopic surgery can be used in patients with focal lesions located in the body or tail of the pancreas.[1] In addition, laparoscopic enucleation of the superficial focal lesion located in the pancreatic head was described; however, in the same study authors recommended to convert to open procedure if the focal lesion is deep in the head of the pancreas.[1] However, major drawbacks were reported regarding the laparoscopic approach, describing the limited tactile feedback to identify a non visible focal lesion with palpation and the inability to use US to evaluate the tiny pancreatic duct.[3] Furthermore, Wen et al.[18] recently reported their successful experience in laparoscopic near-total pancreatic head resection for focal pancreatic head CHI, which was also described as the first case in the literature. In our study, open surgery via a transverse supraumblical laparotomy was performed in all patients. Laparoscopic surgery is recommended for focal lesions located in the tail of the pancreas; however, the limitation of tactile feedback is a major concern in this approach. We believe that palpation of a non visible focal lesion or even a superficial lesion determines the surgical technique in pancreatic surgery. Nevertheless, laparoscopic approach can still be considered a safe option in pancreatic surgery, using additional methods such as ICG that can identify focal lesion and prevent CBD injury.[19]

Bleeding, splenic injury, and CBD injury are the major complications that may occur during pancreatic surgery.[20] Wound infection, prolonged ileus, adhesion obstruction, and wound leakage are also included in postoperative complications.[20] In addition, according to the results of an experienced center, postoperative intussusception after pancreatic resections in children is significantly higher than the general incidence of postoperative intussusception reported in the literature (2.1% *vs.* 0.08-0.25%).[21] Above all, the most significant complications after surgery is recurrent hypoglycemia requiring further pancreatic resection.[20] In terms of complications in our study, we believe that we should have been more careful, particularly in one patient who died due to severe hypoglycemia secondary to persistent vomiting. At that time, we believed that persistent vomiting was not related to surgical causes, and follow-up of this patient was transferred to pediatricians. Nevertheless, we now recognize that surgeons should be more careful when bowel functions do not recover in the optimal time range after surgery or the patient presents with recurrent vomiting after discharge. Since the doses of medication were significantly reduced postoperatively in that patient, we believed that we had removed the focal lesion in the body/tail of the pancreas completely with distal pancreatectomy. However, we recognize that we should have reconsidered further pancreatic resection in that patient. We recommend that surgeons should have a low threshold for performing diagnostic tests in these patients.

There were several limitations to this study. First, due to the retrospective design, the study relied on previous clinical documentations rather than primary data. Second, we could not provide larger sample sizes over the study period due to the rarity of CHI. Lastly, the data was obtained from a single institution, which may limit the generalizability of our outcomes.

In conclusion, the surgical management of CHI is challenging and requires detailed preoperative planning. Although near-total pancreatectomy is preferred in diffuse CHI to avoid severe neurological sequelae due to severe hypoglycemia, it does not provide a complete cure. These patients continue to have hypoglycemia or develop diabetes mellitus and exocrine insufficiency during follow-up. Based on the histologic evidence of higher incidence of focal lesions in pancreatectomy specimens, the main goal should be avoiding unnecessary near-total pancreatectomy in patients with focal CHI and consider pancreas-sparing surgeries in these patients. Knowing the technical details of the surgery, preparation for different surgical approaches to various types of CHI, and being aware of the risky major complications achieve successful outcomes in pancreatic surgery.

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