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Original Article

Pancreatic Operations in Children, an Institutional Experience and Review of Literature

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ABSTRACT

Pancreatic operations are not commonly performed in the pediatric age group. Tumors, chronic pancreatitis, trauma and hyperinsulinemic hypoglycemia are the main diseases leading to pancreatic resections in children. Excluding patients with acute pancreatitis and pseudocysts, over the last 5 years we have performed operations in 11 children for pancreaticoduodenal pathology. The diagnoses encountered were tumors or tumor like conditions (four), angiodysplasia (one), chronic pancreatitis (one), traumatic pancreatic transection (three), traumatic pseudocyst, and neonatal hyperinsulinemic hypoglycemia (one). Operations performed were: pancreaticoduodenectomy (PD) in three patients, subtotal or distal pancreatectomy in six patients, distal pancreaticojejunostomy with oversewing of the proximal pancreatic duct in one patient, and total pancreatectomy, pancreatic cystgastrostomy, and pancreatic biopsies with biliary drainage in a single patient each. There was no operative mortality and one patient developed a pancreatic leak that resolved with drainage. Other morbidity included exocrine (n=3) and endocrine (n=2) insufficiency, intraoperative bile duct injury managed by choledochoduodenostomy (n=1), and delayed gastric emptying (n=1). Adjuvant treatment was needed in two patients for malignant tumors. Overall the children tolerated these pancreatic operations quite well.

INTRODUCTION

The pancreas is not a common site of surgical pathology in children. In children, pancreatic resections are usually performed for neoplasia, chronic pancreatitis (CP), trauma, or persistent hyperinsulinemic hypoglycemia.^{1,2,3,4,5} The main pancreatic tumors in children are pancreatoblastomas, papillary cystic tumors and malignant endocrine tumors of the pancreas.⁶ These are usually managed surgically and evidence for the role of adjuvant therapy is nearly entirely anecdotal. Pancreatic trauma may need surgery for duct disruption or pseudocysts. CP may necessitate a pancreatic ductal drainage procedure or pancreatic resection for intractable pain, biliary complications or when pancreatitis is indistinguishable from a neoplasm.

Children with hyperinsulinemic hypoglycemia who fail medical management require pancreatic resection. Overall, children tolerate pancreatic operations fairly well.

PATIENTS AND METHODS

After obtaining IRB approval a retrospective review was performed of all patients with pancreaticoduodenal pathology requiring pancreatic operations that were less than 18 years of age and were cared for at The Children's Mercy Hospital, Kansas City, MO between 1998 and 2004. Patients with pancreatic pseudocysts secondary to acute pancreatitis were excluded. Eleven patients were identified and reviewed for clinical presentation, operative intervention and outcome with follow-up.

RESULTS

Eleven patients ranging in age from 1 week to 17 years were identified. There were 6 boys and 5 girls. The resections/operations were performed for mass lesions in 4 cases.

CASES 1-4

A 9-year old presented with obstructive jaundice and abdominal pain of brief duration. An abdominal CT scan revealed a mass lesion of the duodenum/pancreatic head (Figure 1). Endoscopic biopsies revealed adenocarcinoma. Preoperative biliary stenting followed by pylorus-sparing pancreaticoduodenectomy (PSPD) was performed. Pathology revealed a poorly differentiated adenocarcinoma with pancreatic and duodenal involvement. Resection margins were negative for malignancy and one of 7 lymph nodes was positive for metastatic adenocarcinoma. Postoperatively there was no evidence of endocrine or exocrine deficiency. Adjuvant therapy with 5 FU and leucovorin was employed and the child remains disease free at 40 months of follow-up.

A 1-week old infant presented with conjugated hyperbilirubinemia. Diagnostic evaluation included an abdominal ultrasound and CT scan that revealed a mass lesion in the head of the pancreas. Pathology after exploratory laparotomy and pancreaticoduodenectomy (PD) revealed infantile myofibromatosis with no evidence of disease in lymph nodes. Pancreatic exocrine deficiency requiring enzymatic replacement developed post-operatively. No adjuvant therapy was required and the child is disease free at 5 years of follow-up.

A 13 year-old was found to have a mass in the region the region of the body and tail of the pancreas on a CT scan performed for abdominal pain. Exploratory laparotomy was undertaken and a distal pancreatectomy was performed for resection of the mass. Final pathologic analysis revealed a solid and cystic papillary epithelial neoplasm with an involved margin of resection. Repeat resection of the margin of the remaining pancreatic tissue along with splenectomy was subsequently performed. Transient hyperglycemia developed after this resection. There was no evidence of disease outside the pancreas and the patient is disease free at 5 years of follow-up.

An 8-year old presented with progressive obstructive jaundice and chronic vague abdominal pain with an abdominal ultrasound and CT scan localizing a mass lesion in the head of the pancreas. There was no associated exocrine deficiency or diabetes. At exploratory laparotomy, multiple transduodenal pancreatic biopsies were performed and revealed only inflammatory changes of chronic pancreatitis. A Roux-en-y hepaticojejunostomy was performed to relieve the biliary obstruction, and the child recovered uneventfully. No adjuvant treatment was needed and the child is doing well at 3 years follow-up with no exocrine or endocrine deficiency.

CASE 5

A 9-year old presented with recurrent upper gastrointestinal bleeds requiring multiple transfusions. An esophagogastroduodenoscopy revealed a vascular malformation of the duodenum and remaining workup was negative. Of note, the patient had undergone a Ladd's procedure for malrotation, and repair of Tetralogy of Fallot as an infant. To accomplish adequate resection, a PD was performed. Postoperatively, a pancreatic leak and lesser sac abscess developed. Operative drainage was performed and the leak resolved. Histology revealed angiodysplasia of the duodenum and non-specific inflammation. The patient has since developed similar lesions in the stomach and proximal small bowel and has required transfusions and endoscopic treatment for the same. Pancreatic exocrine and endocrine deficiency has also developed and enzymatic supplementation and insulin are required at 5 years of follow-up.

CASE 6

A 17-year old presented with long standing, crippling chronic abdominal pain due to chronic pancreatitis with exocrine and endocrine insufficiency. Distal pancreatectomy was performed in an attempt to relieve pain however, was unsuccessful. Five months later the patient underwent completion pancreatectomy. The abdominal pain was relieved after the second operation and pancreatic exocrine and endocrine replacement therapy was continued. Unfortunately the patient died due to unrelated causes 2 years postoperatively.

CASE 7-10

A 9-year old sustained abdominal trauma while riding bicycle and being struck by an automobile, and

a 6-year old presented with abdominal pain after abdominal trauma at play. In both cases, clinical examination revealed epigastric tenderness and abdominal CT scans showed evidence of complete transection of the neck of the pancreas. In both scenarios a spleen preserving distal pancreatectomy was performed within one day of the initial trauma, and their recovery was uneventfully. Neither has developed pancreatic insufficiency in the postoperative period and they are doing well at 4 and 5 years post injury.

A 14-year old presented to the emergency after being a lap belt only restrained passenger involved in a motor vehicle accident. Abdominal CT revealed a pancreatic transection and exploration of the lesser sac revealed a complete pancreatic transection of the neck of the pancreas. The proximal pancreatic duct was ligated and enteric drainage of the distal stump was accomplished via a Roux-en-Y jejunal limb. The postoperative course was uneventful and the patient remains symptom free at 2 years follow-up.

A 16-year old was transferred to our institution 3 days after sustaining abdominal trauma in an MVA. Physical exam findings on admission included peritoneal irritation and fever. A CT scan of the abdomen was obtained and was consistent with pancreatic transection at the level of the body (Figure 2 A-D). Exploratory laparotomy and exploration of the lesser sac was performed and no evidence for pancreatic disruption was identified. External drainage was accomplished via JP drains. These drains were subsequently removed at 2 weeks postoperatively when there had been return of bowel function and appropriate volumes from the drains. Laboratory evaluation for lipase levels of the drain fluid was similar to serum lipase. Two months later a pancreatic pseudocyst developed (Figure 3) that subsequently required cystgastrostomy for obstructive symptoms. The patient remains symptom free while on a low fat diet, however does have recurrent abdominal pain when he eats foods high in fat.

CASE 11

A 3-week old neonate was seen for intractable hypoglycemia despite aggressive medical therapy. A

diagnosis of persistent hyperinsulinemic hypoglycemia was confirmed and a distal (95%) pancreatectomy was performed. Intraoperatively, the distal common bile duct was partially transected near its insertion into the duodenum. This was detected immediately and a choledochoduodenostomy was performed. Subsequently the infant had an uncomplicated postoperative course and pathology revealed presence of focal nesidioblastosis in the pancreas. The patient continues to do well and has not developed diabetes at 4 years of follow up. No adjuvant therapy has been needed. Neurological development is appropriate.

Our patients ranged from 1 week to 17 years of age. Table 1 highlights the clinical data of these patients. These 11 children underwent a multitude of pancreatic operations including: pancreaticoduodenectomy - three (one of them pylorus sparing), subtotal /distal pancreatectomy- six, total pancreatectomy- one, ligation of proximal pancreatic duct and enteric drainage of the distal duct for traumatic transection- one, cystgastrostomy- one, and pancreatic biopsies with biliary drainage- one. Postoperative ileus occurred in all patients (as expected) and mean duration of bowel rest with nasogastric drainage was 5.3 days. Four patients required intraoperative blood transfusions, and there was one bile duct injury that was recognized intraoperatively and managed via choledochoduodenostomy (case 11). This injury was felt to be due to an inappropriately distal insertion of the common bile duct at the junction of the second and third portions of the duodenum. A single patient developed a pancreatic leak leading to an abscess requiring surgical drainage (case 5). One patient had postoperative delayed gastric emptying after a total pancreatectomy (case 6), however she also suffered from long-standing diabetes, which may have contributed to the problem. Postoperative exocrine deficiency was seen in 3 patients and 2 patients developed endocrine deficiency (one was transient). None of our patients developed a pancreatic fistula and there was no operative mortality. Two of the 3 patients with tumors required adjuvant therapy.

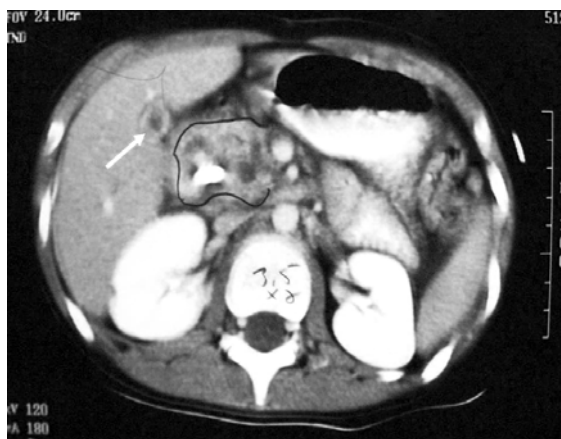


Fig 1. CT scan of the abdomen. The black outline surrounds the large tumor involving the pancreas. The white arrow identifies the obstructed and enlarged common bile duct.



Fig 2. Figures A-D show progressive CT cuts through the body of the pancreas. The arrows identify the location of the suspected pancreatic transection.

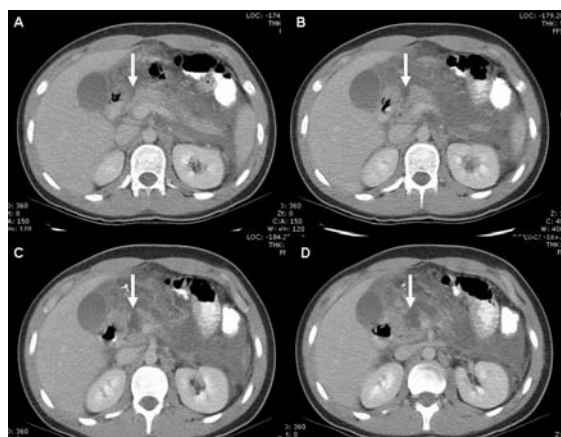


Fig 3. A CT scan of the abdomen at the level of the pancreas and lesser sac. The arrow points out the large pancreatic pseudocyst.

DISCUSSION

Surgical disorders of the pancreas are distinctly uncommon in infancy and childhood.^{2,7} Most pancreatic operations performed in children are due to tumors arising from the pancreas or surrounding tissue or pancreatic trauma.

Pediatric pancreatic tumors arise from exocrine or endocrine tissue and include ductal adenocarcinoma, acinar cell carcinoma, pancreatoblastoma, solid pseudopapillary tumor and endocrine neoplasms. Also, the pancreas may be involved in tumors arising

within the mass of the gland from non-pancreatic cell types adjacent to it and involving the pancreas secondarily. These may be rhabdomyosarcoma,⁸ neuroectodermal tumors,¹ lymphoma,⁹ teratoma¹⁰ and (as in our series) duodenal carcinoma and infantile myofibromatosis.

Pediatric pancreatic tumors often present with abdominal pain and jaundice, which is in contradistinction from the adult population where these findings are uncommon.¹ This likely reflects the fact that tumors are more evenly distributed along the length of the gland in children and ductal adenocarcinoma, the most common pancreatic neoplasm causing jaundice in adults, is extremely rare in children.

Ductal adenocarcinoma of the pancreas has been described in the past, however, Shorter et al,¹ have questioned the diagnosis in the pediatric population. Our patient (case 1), with ductal adenocarcinoma had mucinous and papillary differentiation features that are rarely encountered in ductal adenocarcinoma. Hence, we agree that "classic" ductal adenocarcinoma of the pancreas in the pediatric setting is likely to be an extremely rare tumor.

Pancreatoblastomas are the most common pancreatic tumors seen in childhood.¹¹ These tumors tend to be bulky, solitary and can occur in any region of the pancreas. They contain pluripotent cells on histology,⁶ and their clinical behavior may be very similar to

acinar cell carcinoma.¹ Serum alpha-fetoprotein (AFP) levels are often elevated at diagnosis and may be used to follow these tumors. Surgical resection, whenever feasible is the mainstay of treatment. Chemotherapy (cisplatin, doxorubicin, cyclophosphamide, and etoposide) in the adjuvant and neoadjuvant setting has been used with encouraging results^{1,11,12} and postoperative radiation may have a role for incompletely resected disease.¹³

Solid and papillary epithelial neoplasm of the pancreas is an uncommon low-grade malignant tumor seen predominantly in young females.^{1,11,14} It has been described variously as Frantz's tumor, solid pseudopapillary tumor, papillary cystic neoplasm, solid and papillary neoplasm, and solid and cystic tumor. Generally, this neoplasm presents as an abdominal mass.¹⁴ Focal necrosis, hemorrhage into the tumor and calcification occur commonly,^{1,15} and serum AFP levels are normal.¹⁴ Adequate treatment is complete resection, however local residual disease may not preclude long-term survival.¹ Estrogen and/or progesterone receptors may be present on the tumor cells prompting the use of tamoxifen in its treatment protocol.^{1,16} The role of adjuvant therapy, if any, is not clearly defined.

Acinar carcinoma is an epithelial tumor with evidence of acinar cell differentiation. Its presentation is usually nonspecific and jaundice is infrequent. As many as 16% of patients with acinar carcinoma may present with arthralgias and subcutaneous fat necrosis from elevated serum lipase levels.¹⁷ Effective treatment requires complete excision of the tumor. However, there is a high recurrence rate even after complete resection. Results with chemotherapy and radiation are disappointing.¹⁸ Serous cystadenocarcinoma of the pancreas has not been reported in the pediatric age group.

As in the adult population, insulinoma is the most common endocrine tumor of the pancreas and the majority of them are benign.¹¹ Patients generally present with symptomatic hypoglycemia and elevated insulin levels. Tumor resection is the optimal treatment. Other pancreatic tumors of endocrine origin that may be seen in children are gastrinomas, VIPomas, non-functioning islet cell tumors and mucinous cystic tumors. Johnson and Spitz,¹¹ have recently reviewed these neoplasms.

We treated one child with a duodenal carcinoma presenting as a periampullary lesion. These tumors

are typically seen in the older age group and present with obstructive jaundice. Pathologically, these periampullary tumors arise from the pancreas (62%), ampulla of Vater (19%), distal common bile duct (12%), or duodenum (7%).¹⁹ PD is the standard of care providing the best chance for cure. Extended resections including gastrectomy and/or retroperitoneal lymph node dissection do not improve survival.²⁰ Five-year survival rates for patients with duodenal carcinomas undergoing PD are expected to be greater than 50%.²⁰ Regardless of age, familial adenomatous polyposis predisposes to ampullary neoplasms.²¹

Infantile myofibromatosis is the most common fibrous tumor of infancy.²² Previously known as congenital generalized fibromatosis, it can have solitary or multiple lesions, which may be visceral or peripheral. Infants with visceral lesions usually have a benign course.²² These tumors arise from vascular subintimal mesenchymal or smooth muscle cells and the cellular phenotype is that of myofibroblasts.²³ Surgical resection is the treatment of choice for visceral disease while low dose chemotherapy has been used to manage the generalized form of the disease.^{22,23,24} Spontaneous regression of these tumors is quite common in the peripheral lesions, but extremely rare in the visceral form of the disease.

We performed a PD for angiodysplasia of the duodenum with recurrent life-threatening hemorrhage (case 6). Since, the child has developed similar malformations in the stomach and proximal small bowel. The stomach is the most common site of upper gastrointestinal angiodysplasia, and is asymptomatic in approximately 50% of patients.²⁶ They may, however, present with anemia or relatively large volume UGI bleeds. Endoscopic electrocoagulation or injection sclerotherapy is usually successful in controlling these bleeding episodes. Surgery is indicated only when less invasive therapy is unsuccessful.^{26,27} Aortic stenosis and renal impairment are associated with these lesions.²⁶ Angiodysplasia has also been described in a duodenal diverticulum,²⁸ as well as in the minor papilla.²⁹ Our patient developed a pancreatic leak with an abscess after PD that resolved with drainage. The pancreaticojejunal anastomosis continues to be the "Achilles heel" of the Whipple operation with leak rates ~ 15%.^{30, 31} The leak can generally be managed nonoperatively through careful positioning of intraoperatively placed drains. Pancreaticogastrostomy has been shown to be a safe

and reliable method of reconstruction after PD and may be associated with a lower leak rate than a pancreaticojejunostomy.^{32,33,34} The use of octreotide remains controversial. It continues to be used prophylactically for the prevention of pancreatic leaks after pancreatic resections on the basis of reported studies, primarily in patients with chronic pancreatitis.^{35,36} However, there also exist significant data that show octreotide to be of no benefit in preventing pancreatic leaks and postoperative morbidity after pancreatic resection^{37,38} but rather controls pancreatic volume that may make the leak more manageable. Based on these data we can make no recommendation regarding its use for preventing pancreatic leaks in the pediatric population.

The pancreas and duodenum are the fourth most common injured organs after kidney, spleen and liver in children who sustain blunt abdominal trauma.⁴ Serum amylase and lipase determinations may support clinical suspicion in the diagnosis of pediatric pancreatic trauma but are not reliable or cost-effective as screening tools.³⁹ CT scan remains the main diagnostic modality used in pediatric blunt abdominal trauma. Unfortunately, CT scan may not be conclusive in showing the presence or extent of pancreatic injury.^{3,4} Therefore, in otherwise stable patients with suspected pancreatic ductal injury on initial CT scan, prompt ERCP is recommended.³ Arkowitz et al³⁹ have proposed a useful classification system for pancreatic trauma (Table 2).

A useful algorithm,³ supported by several authors, has been proposed for management of patients with ductal disruption. When significant ductal injury is present, ERCP with stenting should be attempted, followed by stent removal in 10-12 days.^{40,41} If stenting is unsuccessful, distal pancreatectomy is the most commonly performed operation and has very good results with a low morbidity.

Pseudocyst formation and pancreatitis are the most common complications of pancreatic trauma. Approximately 50% of pseudocysts can be managed nonoperatively with the remainder requiring some form of operative drainage.³⁹ A recent study showed that pancreatic leaks after distal pancreatectomy are reduced significantly with ligation of the main pancreatic duct.⁴³

Chronic pancreatitis in childhood is an uncommon but potentially debilitating disorder. Surgery is an option in patients with intractable chronic pain who

fail medical management.⁴⁴ Operations performed include drainage (lateral pancreaticojejunostomy-Puestow procedure) or resectional procedures of varying degree. ERCP has a definite role in the management of this illness as the findings direct further management strategies.⁴⁵ Endoscopic pancreatic drainage and stone removal is a safe and effective modality of treatment for patients with chronic calcifying pancreatitis, in whom elevated intraductal pressures are thought to be the cause of pain.⁴⁶ A longitudinal pancreaticojejunostomy is beneficial for ductal dilatation and associated pseudocysts or pancreatic ascites. However mimicking results in the adult population, the performance of this procedure in the absence of consistent ductal dilatation gives unsatisfactory results.⁴⁷ A resectional procedure may be useful if a Puestow procedure fails to ameliorate pain,⁴⁴ with as many as 75% of patients having a reasonable lifestyle after pancreatic resection.^{48,49} Among resectional procedures, total pancreatectomy has the highest 30-day mortality (5%) and morbidity (47%) and should be avoided. Proximal pancreatectomy will provide pain relief in ~ 90% patients long-term.⁴⁹ Duodenal preserving pancreatic head resection (DPPHR - Beger procedure) is another surgical option that has been recently compared against PD and found to be superior to PD in terms of quality of life, pain relief, nutritional status and length of hospital stay.⁵⁰ Two other surgical options include the Frey procedure (duodenum-preserving resection of the head of the pancreas combined with a longitudinal pancreaticojejunostomy),⁵¹ and duodenum and spleen sparing total pancreatectomy for end-stage CP.⁵² The availability of so many surgical options clearly implies that none of them are ideal procedures or provide consistent results. Regarding deterioration of pancreatic function after pancreatic drainage procedures or partial pancreatectomy for CP, it is not an invariable, immediate consequence of the operation, but rather progression of disease that contributes to this phenomenon.⁵³

Persistent hyperinsulinemic hypoglycemia is the most frequent cause of severe, intractable hypoglycemia in the newborn and infant. Two distinct forms of the disease have been described since 1989: focal and diffuse.⁵⁴ The primary concern in this illness is neurologic injury secondary to severe hypoglycemia. To that end, medical management focuses on maintenance of normoglycemia and consists of dietetic measures, parenteral nutrition, diazoxide,

glucagon, and octreotide. Surgery is indicated only when medical therapy is unsuccessful. Many groups risk of diabetes. Cretolle et al 55 recommend preoperative differentiation between the focal and diffuse form of the disease and favor localization of the focal lesions using pancreatic venous sampling. This can then direct surgical resection with good

treat all patients with near-total or subtotal pancreatectomy, predisposing to a significant success. Despite aggressive treatment, intermediate to severe psychomotor retardation occurs in approximately 20% of these patients with epilepsy affecting a similar number.⁵⁶

Table 1.

CASE #	DIAGNOSIS	OPERATION(S)	OUTCOME
1	Adenocarcinoma	PSPD	Alive/DF
2	Myofibromatosis	PD	Alive/DF/Exocrine Def.
3	Papillary Neoplasm	DP	Alive/DF
4	Chronic Pancreatitis	Hapaticojejunostomy	Alive/DF
5	Vascular Malformation	PD	Alive/Exo & Endocrine Def.
6	Chronic Pancreatitis	DP with delayed CP	Died
7	Pancreatic Transection	SPDP	Alive/DF
8	Pancreatic Transection	SPDP	Alive/DF
9	Pancreatic Transection	Distal Pancreaticojejunostomy	Alive/DF
10	Pancreatic Transection	Pancreatic Cystgastrostomy	Alive//DF/Exocrine Def.
11	Nesidioblastosis	95% Pancreatectomy	Alive/DF

PSPD: Pylorus Sparing Pancreaticoduodenostomy; PD: Pancreaticoduodenostomy; DP: Distal Pancreatectomy; CP: Completion Pancreatectomy; SPDP: Spleen Preserving Distal Pancreatectomy; DF: Disease Free.

Table 2.

Injury Class	Pancreatic Injury
I	Contusion or laceration without ductal injury
II	Distal transection or parenchymal injury with duct injury
III	Proximal transection or parenchymal injury with proximal duct injury
IV	Combined pancreatic and duodenal injury

CONCLUSION

Pancreatic disorders requiring operations are uncommon, but well tolerated in the pediatric population. A multidisciplinary approach, including

dietician, endocrinology, gastroenterology, surgery, and hematology/oncology (for malignant lesions), is necessary in treating these illnesses at diagnosis and over the long-term to provide optimal outcomes. Local complications are the usual cause of morbidity

and pancreatic exocrine and endocrine function needs to be followed postoperatively.

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