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Pancreaticoduodenectomy for malignancies in children

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ABSTRACT

Purpose: Malignant tumors of the common bile duct or of the pancreas head are uncommon in childhood [Perez EA, Gutierrez JC, Koniaris LG, Neville HL, Thompson WR, Sola JE. Malignant pancreatic tumors: incidence and outcome in 58 pediatric patients. J Pediatr Surg. 2009; Jan; 44 (1): 197–203]. With radical surgery being the standard cure for nonmetastatic diseases, pancreaticoduodenectomy (PD) is the best choice when the tumor is localized in the head of the pancreas, or in the lower portion of the common bile duct. The purpose of the present study is to describe five consecutive children managed by PD, and reviewing the particular aspects and results of this rare procedure in children.

Methods: Between 2007 and 2010, five patients (median age: 7 years) underwent PD for nonmetastatic malignant tumors. In two cases, PD was performed en bloc with a right hepatectomy in order to achieve the radical resection of a recurrent biliary sarcoma. Four patients benefited from a "pylorus-preserving" PD procedure. In two patients, resection of the portal vein and vascular reconstruction was performed, and in one case, an extended resection of the biliary ductal system was necessary.

Results: All resection margins were clear. The postoperative course was uneventful, with no pancreatic or biliary leakage in all of the patients. Oral refeeding was achieved by the eighth postoperative day. In two cases, a late revision of pancreatic–jejunal anastomosis was performed because of mild steatorrea and a suspected anastomotic stricture. Two of the patients, who were subsequently operated on second hand, for biliary sarcoma, died from the recurrence; while three of the others, with pancreatic malignancies, are alive and well, with a good functional outcome.

Conclusions: Surgical resection is the treatment of choice for tumors of the pancreatic head area. In the absence of regional or metastatic extension, the radicality of primary intervention is associated with favorable outcomes. Good functionality results were observed after the PD was limited to the head of the pancreas and subject to pylorus-preserving techniques.

In contrast to adult patients in whom the disease is relatively frequent, pancreatic and common bile duct neoplasms are utterly uncommon in the pediatric population. The pancreatic cancer incidence is 0.2% of all pediatric malignancies [1], with among these, pancreatoblastoma being typical of early childhood, while solid or cystic pseudopapillary tumors and neuroendocrine neoplasms can be observed in older groups [2]. In the literature, pancreatic carcinomas and sarcomas are linked to the worst prognosis [2,3].

PD was first performed successfully by Kausch in 1909, and subsequently diffused by Whipple in 1935 [4]. Owing to the rarity of pancreatic cancer in children, it is a procedure that is very rarely performed in this age group. The aim of this retrospective study is to report our experience with PD, its indication, and its results thereof.

1. Patients and methods

1.1. Patients

A retrospective review of the hospital database, for a 4-year study period (2007 to 2010), identified five patients who underwent a PD procedure. Indications were a pancreatic neoplasm in three cases (two pancreatic acinar cell carcinomas, one solid pseudopapillary tumor—ultrasound-guided needle biopsy diagnostic) and rhabdomyosarcoma of the bile ducts (RMSB) in two patients. The latter two patients (Table 1: cases D and E) had been diagnosed, biopsied and initially managed in another center, and they were referred after adjuvant chemotherapy; their locoregional anatomy was complex, and an en bloc DP + partial hepatectomy was considered to be necessary in order to achieve a radical resection (none had metastases, either at diagnosis or preoperatively). In one of these two cases (case D), an explorative laparotomy had been performed at presentation (for biliary obstruction with pseudocystic transformation of the extrahepatic bile ducts), and where a surgical exploration of the "cyst," a biopsy and drainage had been performed: the biopsy

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Table 1
Demographics and characteristics of five children submitted to pancreaticoduodenectomy.

	Gender	Age at surgery (years)	Types of tumor	Surgery	Complications after surgery	Follow-up (months)
Case A	M	7	Pancreatic acinar cell carcinoma (Fig. 3)	Pylorus-sparing PD + portal vein resection and reconstruction	–	26
Case B	F	6.5	Pancreatic acinar cell carcinoma	Pylorus-sparing PD	Stenosis of pancreatic–jejunal anastomosis/resolved	41
Case C	F	12	Solid pseudopapillary tumor	Pylorus-sparing PD	Stenosis of pancreatic–jejunal anastomosis/resolved	24
Case D	F	3	Rhabdomyosarcoma of the extra hepatic biliary tract	Pylorus-sparing PD + right hepatectomy	–	24 (death)
Case E	M	6.5	Rhabdomyosarcoma of the extrahepatic biliary tract (Fig. 2)	PD + conversion to right trisegmentectomy + portal vein reconstruction	–	12 (death)

confirmed the diagnosis of RMSB, but the procedure was further complicated by a chronic biliary fistula to the wound. Although chemotherapy reduced much of the diameter of the tumor mass, the anatomy of the tumor, the previous surgery, and the (likely) neoplastic fistula, would have made it difficult to achieve a radical resection by conventional (limited) resection. The other patient (case E) had previously undergone a right hepatectomy for RMSB in another center, which was followed by a recurrence of the tumor in and around the liver hilum and along the extrahepatic bile duct, which was closely attached to the portal vein trunk.

1.2. Technique

From a technical point of view, the PD was performed according to the classical procedure, as is well described in the literature, but was modified with the preservation of the pylorus when appropriate [4,5] (Fig. 1). The pylorus was preserved when, in the absence of abnormal lymph nodes (at preoperative imaging or at surgery), along the common hepatic artery, a free margin of a minimum of 3 cm was found (macroscopically and during the intervention) between the pylorus and the tumor mass. In two cases, the PD was performed en bloc with the hepatectomy, after the identification and the preservation of the left hepatic artery, and of the portal vein, and without dividing the liver hilum; in one of the two patients (case E), a portion of the portal vein trunk was in continuity with the specimen.

When the resection of the tumor mass had been completed, reconstruction of the digestive system was performed as follows: A Roux jejunal loop was fashioned and positioned through the Treitz hiatus, to reach the supramesocolic area, where it was anastomosed in an end-to-end manner to the pancreatic stump, by separate stitches of

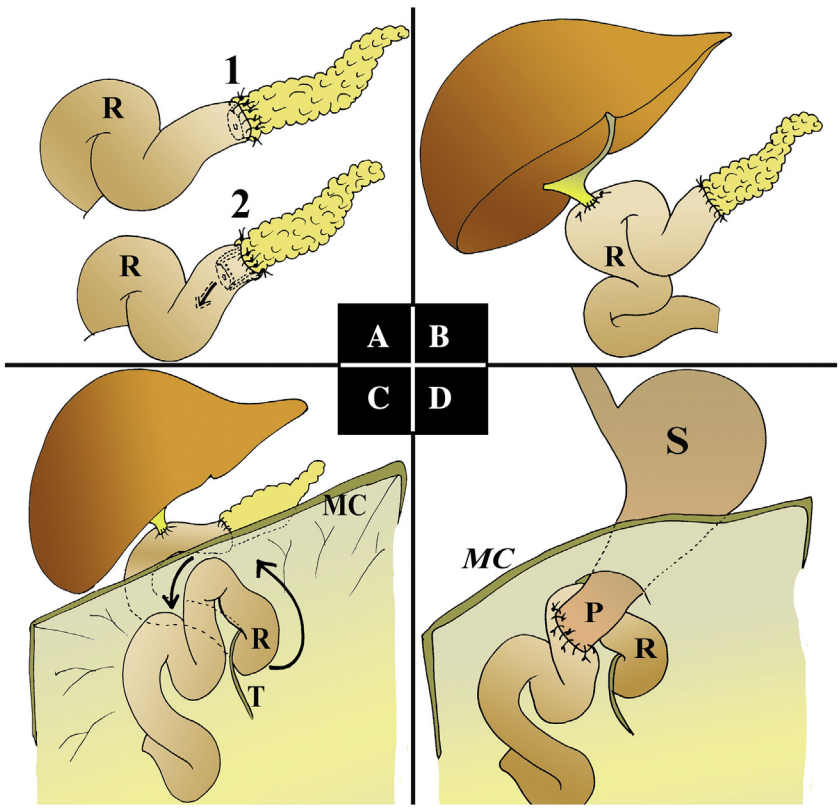

Fig. 1. Technical steps for digestive system reconstruction after pancreaticoduodenectomy (A to D). (A) Pancreaticojejunal end-to-end anastomosis, in double layers (interrupted prolene) with intussusception of the pancreatic stump (2) to cover the first layer (1) (with or without internal stent) (R: Roux-en-Y loop). (B) The hepaticojejunal anastomosis is performed in second, end-to-side, with continuous absorbable monofilament suture and without stent. (C) The 50-cm Jejunal Roux loop has been routed in the supramesocolic area through the Treitz hiatus (T). It is positioned in order to be anastomosed with the gastric outlet (or pylorus) in an isoperistaltic and antireflux (to the Roux) manner. (MC: transverse mesocolon). (D) The gastric outlet—or pylorus (P)—is brought through the mesocolon in order to bring the gastrojejunal anastomosis (continuous absorbable monofilament) within the inframesocolic abdominal area, so that the latter anastomosis is anatomically distant from the previous ones (S: stomach).