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Pancreatectomies for pancreatic neoplasms in pediatric and adolescent age: A single institution experience

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ABSTRACT

Background: There are very few data in the current literature regarding the short- and long-term outcome of surgery for pediatric pancreatic tumors (PPT). No data are available on the impact of pancreatic surgery on the children's growth.
Methods: This is a retrospective cohort study on a consecutive series of pediatric/adolescent patients who underwent pediatric surgery at Karolinska University Hospital from January 2005 to July 2017.
Results: Overall 14 pancreatic operations were performed in 13 patients. The median age was 11.4 years (range 3–15). Six pancreaticoduodenectomies (42.8%), 5 distal pancreatectomies (35.7%), and 3 enucleations (21.5%) were performed. The final histology revealed a solid pseudopapillary tumor in 9 cases (69.2%), neuroblastoma in 1 (7.7%), ganglioneuroma in 1 (7.7%), pancreatoblastoma in 1 (7.7%), and insulinoma in 1 (7.7%). Overall, 3 patients developed post-operative complications (23%). There was no peri-operative mortality. All patients are alive after a median follow-up time of 80 months. Exocrine insufficiency was detected post-operatively in 4 patients (30.7%) Endocrine insufficiency requiring insulin treatment developed in one patient (7.7%). No significant impact on growth was detected in any of the patients after pancreatic resection.
Conclusions: In our series, surgery performed for PPTs seems to be safe and effective. The effect of pancreatic surgery on children's growth does not seem to be significant.

Introduction

Pancreatic tumors in children and adolescents are very rare. Unlike in adults, pancreatic adenocarcinomas are extremely unusual at early age [1]. Pancreatoblastomas are more frequent during the first decade of life, while solid pseudopapillary tumors (SPN) during adolescence [2]. Other, less common tumors, have been reported, too (neuroblastoma, neuroendocrine tumors, acinar cell carcinoma, rhabdomyosarcoma, lymphoma, hemangioendothelioma) [3–5]. Pediatric pancreatic tumors are generally asymptomatic and incidentally discovered during examination for other medical conditions (e.g. trauma). They could also present with non-specific symptoms, such as abdominal pain, vomiting, weight loss or palpable abdominal mass [6]. Irrespective of their histology, pediatric pancreatic tumors have increased in incidence during the last three decades, partially due to the more extensive use of cross-sectional imaging. However, there have not been any significant changes in the therapeutic approach during the same period of time [7]. Surgery remains nowadays the cornerstone of any curative approach, with the only exception of lymphoma [7]. Although the mortality rate after pancreatic surgery performed in highly specialized centers has decreased significantly over the past years, the morbidity rate remains around 30–40% [8]. The vast amount of data on short- and long-term outcome of

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pancreatectomies come from the adult population with pancreatic cancer. Limited information on outcome is available in children and adolescents [3–7,9,10]. The aim of this study is to describe the short-term (mortality and morbidity) and long-term outcome (overall and disease-free survival) of a consecutive series of patients who underwent pancreatic resection for pancreatic tumors during childhood or adolescence at Karolinska University Hospital.

Methods

Study design

This is a retrospective study conducted on a prospectively collected data-base of patients who underwent pancreatic resection at Karolinska University Hospital from 2005 to 2016. An additional patient operated in 1998 at our institution and under follow up was included. All consecutive patients from 0 to 15 years-of-age with surgically treated pancreatic tumors were included. The study was approved by the local ethical committee (2016/2542-31/1).

Pre-operative work-up

All patients were discussed at a dedicated multidisciplinary pancreatic conference. The indication for surgery and/or pre- and post-operative medical oncologic treatment (chemo- and radiotherapy) were discussed with a team of pediatric surgeons and oncologists. Every patient was investigated pre-operatively with a CT-scan and/or MRI. A tumor biopsy was performed in cases where the imaging results were inconclusive.

Measurement of outcome

Patient demographics, clinical presentation, tumor localization, tumor size, and surgical treatment were collected. Intra-operative and peri-operative morbidity and mortality, overall survival, and disease-free survival as well as post-treatment height and weight development were analyzed. Exocrine function was evaluated by the level of fecal elastase. A value of less than 200 μg/g was considered to be indicative of reduced pancreatic exocrine function. Endocrine functional impairment was retrospectively evaluated by the post-operative need for insulin treatment.

Statistics

The overall and disease-free survival were calculated by the Kaplan-Meier method using Graph Pad Prism Software®

Results

Peri-operative results

Overall 14 surgical procedures were performed in 13 pediatric/adolescent patients at Karolinska University Hospital during the study period. Ten of the patients were female (76.9%) and 3 were male (23.1%). The median age was 11.4 years (range 3–15). Patient characteristics are listed in Table 1. Out of the 14 operations performed, 6 were pancreaticoduodenectomies (PD) (42.8%) (Fig. 1), 5 - distal pancreatectomies (DP) (35.7%), and 3 - enucleations (21.5%). One patient, who was initially treated with enucleation, subsequently underwent PD in order to obtain free resection margins (Table 1). The final histology showed a solid pseudopapillary tumor (SPN) in 9 cases (69.2%), neuroblastoma in 1 (7.7%), ganglioneuroma in 1 (7.7%), pancreatoblastoma in 1 (7.7%), and insulinoma in 1 (7.7%). An R0 resection was obtained in 12 patients (92.3%). Pre-operative neo-adjuvant treatment was given to one patient with neuroblastoma (7.7%), while post-operative adjuvant treatment - to 2 patients (15.4%), with neuroblastoma and ganglioneuroma, respectively (Table 1).

Thirteen (92.8%) of the 14 procedures performed were open and 1 (7.2%) was laparoscopic. The median operative time was 4.7 h (range 1.5–16 h). The median blood loss was 144 ml (range 10–500 ml). The median hospital stay was 16.4 days (range 8–40). The longest post-operative stay was related to the decision to perform a PD after a non-radical enucleation of a SPN. Overall, 3 patients developed post-operative complications (23%). In 2 of them (15.4%) the complication severity was ≥ Clavien 3 [11]. A post-operative pancreatic fistula developed after 2 of the 14 operations (14.3%) - one biochemical leak and one grade B [12]. Gastro-intestinal bleeding was observed in one case (7.7%) after a PD, related to the pancytopenia that was induced by the neo-adjuvant treatment. All complications were treated conservatively and there were no post-operative reoperations or death.

Endocrine and exocrine function

Data regarding pancreatic exocrine and endocrine function were available on all patients included in the study. Exocrine insufficiency was detected post-operatively in 4 patients (30.7%) and corrected by administration of pancreatic enzymes. Three of these patients (75%) underwent pancreaticoduodenectomy and one (25%) - distal pancreatectomy. Endocrine insufficiency requiring insulin treatment developed in one patient (7.7%) who underwent distal pancreatectomy.

Long term outcome and growth

All 13 patients who underwent pancreatic surgery for pancreatic neoplasms are alive after a median follow-up time of 80 months (range 2–239 months). One of the 13 patients was diagnosed with intracranial metastases 6 months after the resection for abdominal neuroblastoma infiltrating the head of the pancreas. There were no local recurrences detected in this group of patients. (Fig. 2).

Growth after surgery was evaluated by estimating the expected weight and height development individually for each patient based on pre-treatment growth charts [13]. In this series of 13 patients, we identified 6 patients who had a significant remaining growth potential at the time of surgery, as well as a sufficient follow-up time for determination of post-treatment height development. Relevant data on height development were available in 5 of these cases (age range 4–12 years) (Table 1, patients: 3, 6, 8, 10 and 13). Five patients who had already reached their final height at the time of surgery, as indicated by a decreased growth rate on the growth curves, and two patients in whom the follow-up time was insufficient to draw conclusions about the effect of pancreatic surgery on height development, were excluded from this analysis. The median follow-up with regard to height development was 80 months (range 9–159 months). The median height at the last follow-up visit was at −0,5 SD (Range: +0,5 to −1,5 SD). Four of five patients followed their expected growth curves after surgery. One patient deviated to slightly shorter (−1,5 SD) final height than the estimate, based on the length of his parents (−0,5 SD) (Table 1).

In 11 out of 13 patients, the follow-up time was sufficient to determine the post-treatment weight development and relevant data was available in 10 of these cases. The median follow-up time was 102 months (range 9–159 months). The median recorded weight at the last follow up visit was at −0,5 SD (range: +1,5 SD to − 2,0 SD) (Table 1).

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