Pancreaticoduodenectomy in Children: Optimising Outcome of Uncommon Paediatric Procedures

Abstract:
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Abstract
Contemporary surgical practice is increasingly dominated by subspecialisation in response to improved outcome from high volume centres, though uncertainties persist for uncommon paediatric procedures. Three paediatric pancreaticoduodenectomies performed at Our Lady’s Children’s Hospital, Dublin, over a period of 9 years were evaluated to substantiate their continuing performance by paediatric rather than adult pancreatic surgeons. With ages ranging from 18 months to 8 years old, the mean operating time was 263 minutes, while the average hospital stay was 12 days. There was no perioperative mortality, although complication rate was 100%. Re-operation was required in 33%. The long term outcome of this small paediatric cohort was comparable to adult series despite the low patient accrual, underscoring the advantages of a multidisciplinary approach afforded by tertiary paediatric institutions for intricate yet infrequent operations in children.

Introduction
The era of subspecialisation has consigned paediatric surgery to one of the last bastions for the general surgeon. Widely embraced for improving outcome through the integration of expertise and resources, regionalisation of services is firmly established even within paediatric surgery for procedures including appendectomy and pyloromyotomy. Yet, uncertainties persist for uncommon procedures of rare paediatric disorders. A case in point is pancreaticoduodenectomy, an operation notorious for its high morbidity and mortality. Adult pancreaticoduodenectomy remains a complex, high-risk procedure, refined over decades to one of satisfactory outcome as deemed by its current mortality rate of less than 5%. Crucial to this successful evolution is the advantageous relationship between high patient volume and good outcome. The occasional need for this procedure in children can therefore be intimidating given the dearth of large paediatric series. Nonetheless, paediatric surgeons are expected to be proficient in all procedures performed in newborns and children, a scenario particularly embellished in developing countries where the rising young population overwhelms the number of new surgeons. The aim of this study was therefore to examine if paediatric surgeons should continue to perform paediatric pancreaticoduodenectomies, and to explore strategies to optimise their outcome within the context of a tertiary paediatric institution in Ireland.

Methods
Pancreaticoduodenectomies for various indications at Our Lady’s Children’s Hospital were retrospectively analysed. Data was extracted using the Hospital In-Patient Enquiry (HIPE) system. With reference to adult series, operative technicalities, complications, morbidity and mortality were some of the parameters scrutinised.

Results
Between January 2000 and December 2009, three patients underwent pancreaticoduodenectomies. A 4-year old boy presented with obstructive jaundice secondary to a pancreatic head mass. Although trucut biopsy revealed only chronic inflammation, worsening biliary obstruction necessitated a bypass procedure (choledochoduodenotomy). Progressive increase of the mass, recurrent upper abdominal pain and lingering doubts of malignancy mandated its excision. Complete resection left approximately 60% of total pancreatic volume, resulting in pancreatic insufficiency within the next few months. Pancreatic enzyme and vitamins replacement therapies were administered and gradually discontinued after three years. The final histology revealed chronic pancreatitis related to malformation of the pancreatico-biliary ductal system.

Figure 1: Large PNET posterior to head of pancreas

An 18-month old girl presented with massive upper gastrointestinal haemorrhage and uncompensated anaemia secondary to a large eroding duodenal ulcer. Despite ulcer under-running, haemorrhage recurred. Re-laparotomy revealed erosion into the gastroduodenal artery and adjacent pancreas. Successful haemostasis was only achieved following a limited resection of the head of pancreas. Zollinger-Ellison syndrome was later excluded, and histology supported the diagnosis of stress ulceration. Early postoperative period was complicated by lymphatic leak, which subsided after two weeks of parenteral nutrition and peritoneal drainage. Surveillance endoscopy revealed no recurrent ulceration.

Figure 1: Large PNET posterior to head of pancreas
An 8-year-old girl presented with obstructive jaundice, abdominal pain, leucone and an upper abdominal mass. CT scan revealed a large retroperitoneal tumour adjacent to the duodenum. After histological confirmation of a primitive neuroectodermal tumour (PNET), down staging chemotherapy was commenced. Tumour progression despite chemotherapy was palliated with percutaneous biliary drainage. An alternative approach yielded the desired response. The pancreatic tumour with its duodenal infiltration was completely excised a month later. Histology confirmed a stage 3 PNET with extensive vascular invasion. Recovery was further complicated by ischemic stricture of the gastro-duodenal anastomosis, which was readily revised. Surveillance CT scan at one year showed no recurrence. All reconstruction was via a pylorus-preserving pancreaticoduodenectomy. Operating time ranged from 174 to 330 (mean, 263) minutes. No intraoperative blood transfusion was required. Hospital stays were between 9 to 17 days, of which 2 to 4 days were spent in ICU. All underwent contrast study within first week of surgery to confirm anastomotic integrity. Subsequent follow ups ranged from 1 to 9 years, with no recurrence.

Figure 2: Stricture of gastro-duodenal anastomosis secondary to ischaemia (arrows)

Discussion

The uniqueness of this series lies in the application of pancreaticoduodenectomy for a diverse range of pancreatic disorders in children, as opposed to its usual oncological indication. Clearly, this operation remains an invaluable armamentarium in the management of severe duodenal haemorrhage, chronic pancreatic fibrosis and pancreatic tumour, which were all satisfactorily treated without long term sequelae. In all cases the pylorus was preserved without compromising margin, a modified technique with better outcome than when the pylorus is sacrificed. Time, hospital stay, transfusion requirement and operative mortality were comparable to paediatric counterparts. However, complications consisting of pancreatic insufficiency, lymphatic leak and anastomotic stricture were present in all cases, versus a 41% complication rate in adult patients. In the absence of patient or operative complexities, it would appear futile to surmise if the outcome would be improved by a larger series.

Questions linger as case volume is unlikely to increase substantially as far as paediatric pancreaticoduodenectomy is concerned. This low patient anecdotal diminish the significance of comparing paediatric and adult series. With only three cases spanning nine years, it would be difficult even for tertiary institutions such as ours to achieve the status of a high volume centre, defined as one that performs more than five cases annually. Continuing performance of pancreaticoduodenectomy by paediatric surgeons may lie in the transfer of expertise gained from adults to children. In institutions accustomed to dealing with a large volume of index cases, paediatric surgeons should be able to execute this transferance with ease. Ideally, pancreaticoduodenectomy in children should be performed by paediatric surgeons with advanced training in liver and pancreatic surgery, as is the policy of our institution.

Additionally, several factors may be crucial to ensure an optimal outcome. Firstly, most paediatric surgeons with foundation training in adult general surgery would have been exposed to and have even performed these complex procedures in the course of their training. The procedure can be readily familiarised from a joint operation of pancreatic and paediatric surgeons. The pancreatic surgeons technical proficiency can be effectively married with the paediatric surgeons in depth knowledge of paediatric anatomy and physiology. Collaboration is also invaluable for the expertise of endoscopic stenting for the more complicated cases. A child's excellent capacity for healing, superior nutritional status and lack of co-morbidities coupled with the more favourable disease pathology contribute to this better outcome. Another factor of critical importance is the provision of specialised post operative care including skilled nursing and paediatric intensive care, supported by specialties such as oncology, gastroenterology and dietetics that would corroborate the overall management for pancreaticoduodenectomy in a paediatric rather than an adult hospital. In short, we advocate a multidisciplinary approach to challenging operations such as pancreaticoduodenectomy with a setting of a major paediatric institution. Despite rarely performed, careful patient preparation, meticulous operative techniques and comprehensive post operative care should warrant an optimal outcome.

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References