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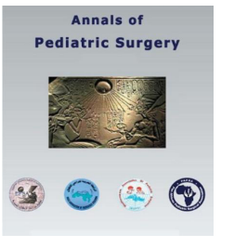

Case Reports

Cephalic Duodenopancreatectomy In The Neonatal Period: A Case Report And Review Of The Literature

Bada-Bosch I[1], Cerdá J [1], De La Torre M [1], Sánchez Sánchez C[2], Tolín Hernani M[2], Miranda Cid C[2], Del Cañizo A[1], Molina E [1], De Agustín JC[1].

[1] Department of Pediatric Surgery., [2] Department of Pediatric Gastroenterology, Hospital Universitario Gregorio Marañón. Calle O`Donnell 48-50, 28009, Madrid, Spain..

ABSTRACT

Background: Pancreatic masses in the neonatal period are a rare pathology. Resection of the head of the pancreas is an exceptional surgical challenge due to the need to perform millimetric anastomosis, therefore, subsequent complications are frequent.

Methods: Case report and review of published articles on cephalic duodenopancreatectomies (CDP) in patients less than 28 days-old.

Case report: An 11-days-old female newborn, 3030g, consulted for apnea and cyanosis. Magnetic resonance imaging showed a cystic mass in the right hypochondrium measuring 6.2x5.9x6.4mm with suspicion of malignancy. At 22 days of life a laparotomy was performed finding a mass in the head of the pancreas firmly adhered to the extrahepatic biliary tract and duodenum. A CDP and reconstruction by gastroduodenostomy, hepatico-duodenostomy (Kasai-like) and dunking pancreato-gastrostomy were performed. The pathological anatomy described a serous cystadenoma. With 6 years of follow-up, she has presented a single episode of cholangitis, with no signs of endocrine or exocrine insufficiency.

Results (Table 1): 6 patients with CDP less than 28 days have been described. The surgical technique presented many variations. Five patients presented complications, the most frequent being exocrine insufficiency.

Conclusion: CDP in the neonatal period is an exceptional procedure, with only 5 other cases described in the literature. We present CDP on the largest mass described so far, with no signs of pancreatic insufficiency in a long-term follow-up.

Key words: Cephalic duodenopancreatectomy, neonatal, pancreatic mass ,Whipple procedure.

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INTRODUCTION

Cephalic duodenopancreatectomy(CPD) or Whipple procedure is a surgical procedure that consists of resection of the head of the pancreas and duodenum. It can also include the resection of the biliary tract (partially or completely), the distal stomach and first loop of jejunum. It is the most

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Corresponding Author: sabel Bada Bosch. MD. Hospital Universitario Gregorio Marañón. Pediatric Surgery Department., Calle O`Donnell 48-50, 28009, Madrid, Spain **, Tel.: +34 915290039: E-mail: isabel.bada.bosch@gmail.com**

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common surgical treatment for pancreatic head tumors in adults. However, it is an extremely infrequent surgery in the pediatric population due to the rarity of childhood pancreatic tumors, being exceptional in the neonatal period.

We present a case of a patient who required a CPD for a pancreatic tumor with suspected malignancy. Due to the rarity of the procedure, a review of the literature on neonatal CPD has also been performed.

METHODS

Description of a clinical case. Written informed consent was obtained from the legal guardians for publication of this case report and accompanying images. No identifiable images appear in this manuscript.

Subsequently a literature review was performed in Pubmed in January 2023. The keywords used were: "Whipple procedure" [OR] "cephalic duodenopancreatectomy" [AND] "neonatal" [OR] "pediatric". From the results obtained, abstracts were reviewed and articles describing at least one case of CPD in a patient less than 28 days old were selected for full revision.

CASE REPORT

An 11-day-old newborn (3030g) was admitted to the emergency department for apnea and cyanosis episodes. She had had an uneventful pregnancy and delivery with normal prenatal ultrasounds. No signs of irritability, vomiting or jaundice were present. On examination she presented mild hepatomegaly and a firm mass in the right hypochondrium.

An abdominal ultrasound was performed (Figure1) identifying a microcystic mass in the right hypochondrium measuring 64x51x59mm, without clear dependence of any organ. The liver parenchyma and biliary tract were normal. The pancreas could not be identified. She also had a right pyelocaliceal dilatation. A magnetic resonance was performed (Figure 2) confirming a multicystic mass of 62x59x64 mm (volume 117cc). It was hyperintense in T2-weighted images and hypointense in T1. Again, dependence of a particular organ could not be defined but radiological characteristics such as compression of the inferior vena cava, right renal vein and ipsilateral pyelocaliceal dilatation were evocative of a malignant mass, therefore surgical resection was advised.

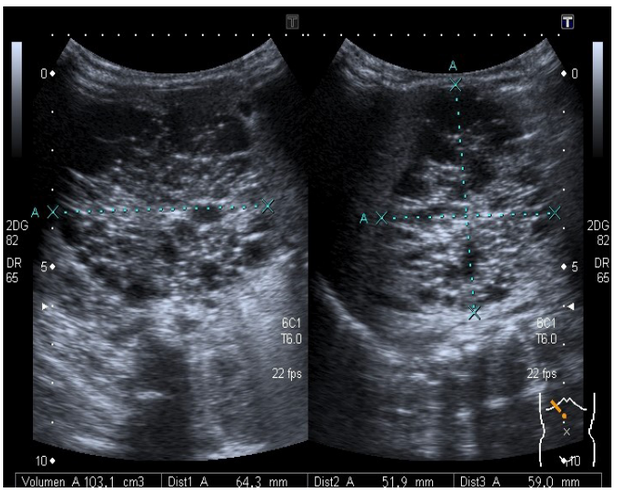

Fig1. Abdominal ultrasound. A microcystic mass of 103cc is observed.

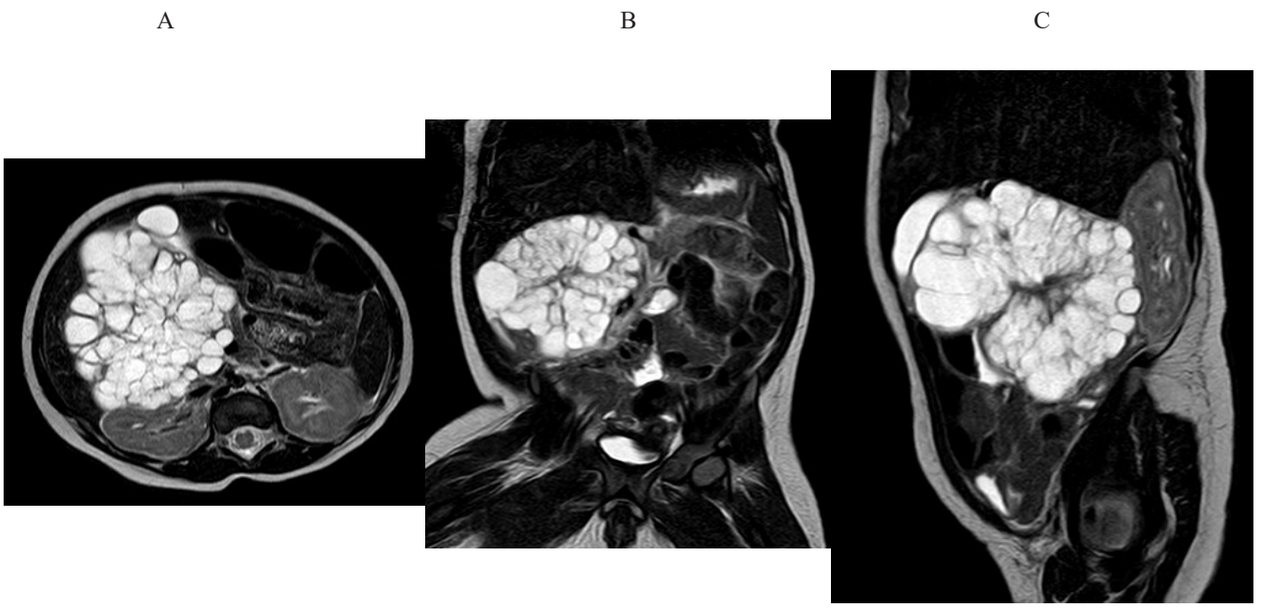

Fig: 2. Abdominal MRI, T2 weighted images. Axial (A), coronal (B) and sagittal (C) views. They show a well-delimited hyperintense multicystic mass with anteroposterior diameter of 62 mm, longitudinal 59 mm and transverse 64 mm. In the center there is a hypointense solid stellate area. It produces compression of the inferior vena cava, renal vein and right kidney. No pancreatic head is seen in any of the sections.

A complete work-up was conducted, including blood analysis (complete blood count, liver and renal function panel, and coagulation tests, alpha-fetoprotein), blood culture, urine culture, respiratory virus panel, electrocardiogram, echocardiogram, transfontanel ultrasound, and cerebral monitoring, all of which yielded normal results.

At 22 days of life a laparotomy was performed finding a mass dependent of the head of the pancreas. Its size and close relationship to surrounding structures made its enucleation impossible following oncological principles. It was firmly adhered to the extrahepatic biliary tract and the first and second portions of the duodenum, we decided to perform a CPD (Figure 3).

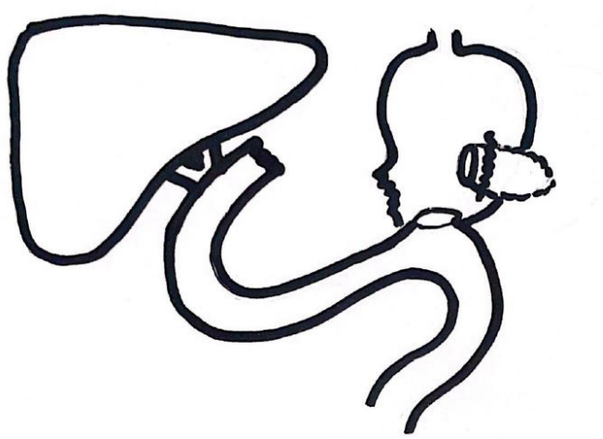

Fig. 3. Surgical scheme. Resection of distal stomach, 1st and 2nd duodenal portion, pancreatic head and biliary tract. Reconstruction with hepatico-duodenostomy, pancreato-gastrostomy and gastro-jejunostomy.

Intraoperative cholangiography showed a lesion of the common bile duct; thus, cholecystectomy and resection of the extrahepatic bile tract were indicated. The common hepatic duct was filiform (less than a millimeter) which made a regular hepatico-duodenostomy a high-risk anastomosis. For this reason, we performed a Kasai-like anastomosis to the biliary plate instead. Pancreatic section was achieved with an endo-GIA between the body and tail of the pancreas respecting tumor-free margins. The technical difficulty in freeing the tail of the pancreas prevented from a duodenal anastomosis, so a dunking pancreato-gastrostomy was performed. For this purpose, the anterior aspect of the stomach was opened at the greater curvature and a posterior wall incision was achieved through the anterior opening. The tail of the pancreas was introduced into the stomach through the posterior incision and marsupialized with a purse string suture. Gastrointestinal continuity was achieved by means of a side-to-side gastro-jejunostomy anastomosed at 17cm from the hepatico-duodenostomy. Two drains were left in the hepatico-duodenostomy's bed.

Postoperative period presented no notable surgical complications except for a persistent serous drainage (with normal amylase and bilirubin levels) through one of the drainage wounds. The patient was discharged 2 months after the surgery. The pathological anatomy described a massed composed of thin-walled cysts with flat or cuboidal epithelium surrounded by normal pancreatic tissue, therefore a serous cystadenoma of the pancreas with free margins.

During follow-up she has been diagnosed with KBG syndrome secondary to a mutation in the ANKRD11 gene. It includes dysmorphic features, short stature, mild psychomotor retardation and renal cysts but no pancreatic masses have been described. During the 6 years of follow-up, she has presented a single episode of cholangitis at 2 years of age and occasional episodes of late dumping symptomatology, currently resolved. She presents analytical criteria of mild exocrine pancreatic insufficiency (minimal elevation of fecal elastase) without steatorrhea or gastrointestinal symptoms and normal glycemic controls. She is under prophylactic treatment for cholangitis with ursodeoxycholic acid and sulfamethoxazole. She has not presented signs of recurrence of the pathology.

RESULTS (TABLE 1)

Eight articles describe cases of CPD in the neonatal period[1–7], including the one presented. In these 8 articles 6 patients are described. After full analysis we found that one of the patients described in Jaksic et al in 1992[3] corresponds to the case reported by Rich et al in 1986[2]. The same is true for the article by Leung et al in 2014[5] and the one by Park et al in 2016[7].

Four males and two females have been described with a minimum age of 5 days and maximum of 2 months (first and second surgeries being at 24h and 12 days). None of the patients had prenatal diagnosis of the mass, all presented symptoms: 2 bilious emesis, 2 jaundice, 1 low platelet count, 1 symptomatic hypoglycemia and 1 apnea episodes.

All patients underwent ultrasound for the diagnosis of the mass, which was later completed with computed tomography in 4, magnetic resonance in 1 and a contrast study in 1. The size of the mass comprised between 2 and 6.5cm (the case described above).

Regarding the surgical procedure, in all cases the head of the pancreas and the duodenum were partially or totally removed; the distal stomach was resected in 3 patients, the common bile duct in 3 and the gallbladder in 3. The reconstruction of the intestinal transit was performed via a gastro-jejunostomy in 4 patients and duodeno-jejunostomy in 1. Biliary anastomosis was performed as common bile duct-jejunostomy in 2, Kasai-like hepatico-jejunostomy in 2 and conventional hepatico-jejunostomy in 1. Pancreatic reconstruction was performed by pancreatic-jejunostomy in 3, pancreatic-gastrostomy in 1 and pancreatic isolation was performed in 1 patient. Jiao et al (6) do not describe the exact surgical technique.

Table 1. Neonatal CPD reported in literature

Authors	Age at surgery	Symptoms	Imaging	Size	Excision	Reconstruction	Pathological Anatomy	Complications
Shearburn 1975[1]	♀ 2 months	Bilious emesis	NS	2x2cm	Head of the pancreas Duodenum Distal stomach Distal CBD	Gastro-jejunostomy EE Coledocho-jejunostomy ES Pancreas oversewn Gastrostomy Truncal vaguectomy	Fibrosarcoma	Exocrine insufficiency Endocrine insufficiency (self-limited) Growth delay
Rich 1986[2]	♂ 21 days	Jaundice	Ultrasound CT	6cm	Head of the pancreas Duodenum CBD Gallbladder	Duodeno-jejunostomy Hepatico-jejunostomy (Kasai-like) Pancreatico-jejunostomy	Pancreatoblastoma	Exocrine insufficiency Colangitis for 2 years Hepatico-jejunostomy anastomotic stricture, secondary cirrhosis
Morrow 1999[4]	♂ 5 days	Jaundice	Ultrasound CT	2cm	Head of the pancreas Distal stomach Duodenum Distal CBD	Gastro-jejunostomy Coledoco-jejunostomy ES Pancreatico-jejunostomy EE	Neonatal myofibromatosis	Exocrine insufficiency (self-limited)
Leung 2014[5]	♂ 7 days	Bilious emesis Low platelet count	Ultrasound UGIC CT	4x4x5cm	Head of the pancreas Distal stomach Duodenum 15cm jejunum CBD Gallbladder	Gastro-jejunostomy Hepatico-jejunostomy Pancreatico-jejunostomy dunking	Kaposiform hemangioendo-thelioma	Exocrine insufficiency
Jiao 2015[6]	♀ NS	Symptomatic hypoglycaemia	Ultrasound CT	2.2 x 3.7 x 3.9cm	NS	NS	Intraductal papillary mucinous neoplasm	NO
Bada-Bosch 2023	♀ 22 days	Apnea Cyanosis	Ultrasound MRI	6.2 x 5.9 x 6.4 cm	Head of the pancreas 1st-2nd part of duodenum CBD Gallbladder	Gastro-jejunostomy SS Hepatico-duodenostomy (Kasai-like) Pancreatico-gastrostomy dunking	Serous cystadenoma	Dumping (self-limited) 1 episode of cholangitis Growth delay (associated syndrome)

Abbreviations. NS: non specified. CBD: common biliary duct. CT: computerised tomography. UGIC: upper gastrointestinal contrast. MRI: Magnetic Resonance Imaging. ES: end-to-side. EE: end-to-end. SS: side-to-side

The pathological anatomy described three malignant tumors (fibrosarcoma, pancreatoblastoma, kaposiform hemangioendothelioma), 1 borderline lesion (papillary intraductal mucinous neoplasm) and 2 benign masses (infantile myofibromatosis, serous cystadenoma).

None of the cases described surgical complications in the immediate postoperative period. In the long-term follow-up all but one patient[6] (83.3%) report some type of sequelae, most of them mild. The most frequent complication is exocrine insufficiency in 4 (66.7%) requiring chronic enzymatic treatment in 2, dietary adaptations in 1 no treatment (self-limited) in 1 patient. One patient had transient endocrine insufficiency requiring insulin treatment. Two patients presented with staturo-ponderal delay. Two other patients had cholangitis, one of them multiple episodes but currently resolved and another patient had a single episode. The only severe complication in the cohort was in the patient reported by Rich et al[2] who presented a stenosis of the biliary anastomosis causing liver cirrhosis that is pending transplantation at the time of publication of the article by Jaksic et al[3].

DISCUSSION

Pancreatic tumors have an estimated incidence of 0.191 per million inhabitants in children under the age of 19 (8,9). They account for only 0.2% of pediatric neoplasms(10) and 4% of rare tumors(8). Their incidence has not been described in the neonatal period. The most frequent histology is that of pseudopapillary neoplasm, however this is more frequent in adolescent patients, while pancreatoblastoma is the predominant histology in infants.

With the exception of lymphomas, pancreatic tumors always have surgical indication. The type of surgery depends mainly on the location of the tumor. Since pancreatic tumors have a better prognosis in children than in adults, aggressive surgeries should be avoided as far as possible[3]. However, due to the characteristics of the mass in terms of size and relationship to neighboring organs this is not always possible respecting the principles of oncological surgery. The location in the head of the pancreas accounts for slightly less than 50% of tumors, and surgery in these patients is more complex. In a review of 104 pediatric patients with pancreatic tumors[10], 43 patients had the tumor in the head of the pancreas requiring CPD in 23% and resection of the head with duodenal preservation in 58%, enucleation being possible in only 18.6%.

CPD is a technically demanding procedure even in adult patients due to the need to respect neighboring vital structures and the numerous high-risk anastomoses. In children, and especially in neonates, this becomes an exceptional surgical challenge owing to the millimetric size of the anatomical structures. Due to these technical difficulties, CPD, standardized in adults (Whipple technique with Child type reconstruction), presents many variations in children, as we have seen in the cases presented in the literature. The organs being anastomosed and the type of anastomosis depended more on the proximity and disposition of the structures than on the theoretical surgical technique. This means that the pediatric surgeon must have not only sufficient skill to perform millimetric anastomoses, but also a wide knowledge of surgical variations and different types of anastomoses depending on the patient's anatomy and the mass. In our case, preservation of the common bile duct was not possible because it was damaged by the compression of the mass, nor was it possible to preserve the common hepatic duct because of its filiform caliber, that prevented a safe anastomosis. Therefore, the Kasai-like anastomosis was chosen, which is the most common procedure in pediatric surgery for the treatment of biliary atresia. Regarding the pancreatic anastomosis, the disposition of the body and tail of the pancreas made it difficult to perform a duodenal anastomosis, which would have remained under tension. With a limited resection of the gastric antrum, the greater curvature of the stomach was anatomically closer. Again, because of the caliber, it was impossible to perform a Wirsung-gastric anastomosis safely as in adults, so an invaginating dunking anastomosis was chosen on the posterior gastric wall. Although the advantages of pyloric preservation are well known, due to the large size of the mass in our patient, it was not compatible with an adequate oncological resection.

Despite the great technical difficulty of the procedure, neither our patient nor any of the patients described in the literature presented immediate postoperative complications such as those described in adult population (pancreatic fistula, biliary leak...). Although this is a small series, it is described in the literature that CPD in patients under 30 years of age has a significantly lower rate of complications than in those over 30 years of age (29.4% vs 40%)[9]. This lower rate of complications could be due to a better general and nutritional status compared to adult patients with malnutrition in relation to pancreatic carcinoma and more comorbidities. In a larger cohort of pediatric patients (104 patients, 43 pancreatic head tumors) Cheng et al[10] described 37.2% of pancreatic fistulae, 7.5% of biliary leaks, 23.2% intra-abdominal infections and 13% of pancreatectomy hemorrhages.

Regarding long-term complications, only one of the patients presented a severe complication due to biliary anastomosis stenosis that went unnoticed. Exocrine pancreatic insufficiency is one of the most frequent complications in adult (56-73%) and children (14.6%) series[10]. Similarly, a high rate of patients operated on in the neonatal period present to some degree exocrine insufficiency, probably due to the difficulty of preserving healthy pancreatic parenchyma in a neonatal size pancreas. As for endocrine insufficiency, we have not found any patients operated on in the neonatal period who present it in the long term, compared to a rate of 20-25% in adults[10]. This is probably due to a follow-up that is too short in time, together with the fact of a basal healthy remanent pancreatic tissue as opposed to the chronic fibrosis present in most adults with pancreatic carcinoma. Therefore, most pediatric patients who undergo surgery for CPD will have an adequate quality

of life as long as no major complications occur[7].

CONCLUSION

Neonatal pancreatic tumors are exceptional and always surgical. The most common procedure for the treatment of those located in the head of the pancreas is CPD. It is a surgery of high technical complexity and with many variations in terms of reconstruction. Nevertheless, we present the case of a patient with an adequate evolution and long-term follow-up

DECLARATIONS

Ethics approval: no ethics approval was needed for a case report Publication.

Consent for publication: Written informed consent was obtained from the legal guardians for publication of this case report and accompanying images. No identifiable images appear in this manuscript.

CONSENT FOR PUBLICATION

Not applicable.

AVAILABILITY OF DATA AND MATERIAL

Not applicable.

COMPETING INTERESTS

The authors declare that they have no competing interests

FUNDING

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AUTHORS' CONTRIBUTIONS

• IBB designed the first draft of the paper and subsequent corrections and performed the literature review.

• JAC is the main surgeon responsible for the patient. He reported on clinical information and contributed on the design of the first draft.

• MT performed the literature review.

• CSS, MMTH and CM are the gastroenterologist responsible for the long-term follow-up of this patient. They contributed to the description of the case.

• AC, EM and JCA are part of the surgical team responsible for the initial management and follow-up of the patient. They contributed to the description of the case.

• All authors have reviewed and approved the final version of the manuscript.

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Not applicable.

ABBREVIATIONS

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