Paediatric Abdominal Oncovascular Surgery: a Single-centre Experience and Review of the Literature

Andrea Zanini (✉ andrea.zanini.87@gmail.com)

Department of Paediatric Surgery, Faculty of Health Sciences, University of Witwatersrand, Johannesburg, South Africa

Derek Harrison

Department of Paediatric Surgery, Faculty of Health Sciences, University of Witwatersrand, Johannesburg, South Africa

Sean Connor

Department of Paediatric Surgery, Faculty of Health Sciences, University of Witwatersrand, Johannesburg, South Africa

Jerome Loveland

Department of Paediatric Surgery, Faculty of Health Sciences, University of Witwatersrand, Johannesburg, South Africa

Abstract

PURPOSE Oncovascular surgery is a term identifying vascular resection in the context of tumour resection which is still controversial in children. We present our experience and review of the literature.

METHODS A retrospective review of children who underwent abdominal oncovascular procedures in our Institution from 2018 to 2022 was conducted. Type of operation and post-operative outcome was described. Literature review on oncovascular surgery in children is presented.

RESULTS Seven cases were identified, mean age 8.25 years (9 months- 14 years): 2 bilateral paraganglioma, 2 Wilms tumour, 1 Pancreatoblastoma, 1 Solid Pseudopapillary Tumour of the Pancreas (SPN), and 1 Hepatoblastoma. Five procedures were performed on the Inferior Vena Cava: 3 patients underwent cavectomy with no reconstruction, one had a partial cavectomy with primary repair, and one had resection and reconstruction with a PTFE graft which complicated with leaking and infection. Two patients underwent a pancreaticoduodenectomy with Portal Vein resection and primary anastomosis. Five patients were completely resected (R0), 2 patients had microscopically positive margins (R1). All patients were alive at last follow up.

CONCLUSION Vascular resection can allow complete tumour resection in locally advanced paediatric tumours. Oncovascular surgery in children is feasible and may be beneficial in selected cases.

Introduction

Although commonly complicating certain adult solid tumours, vascular involvement in paediatric oncology is rare.[1] While vascular invasion by tumour has traditionally been considered a contraindication for resectability, over the past decades, resection of the vascular component concomitant with resection of the primary tumour have been reported, extending the boundaries of tumour resectability in selected cases.[1,2] While in adult surgical oncology some of these vascular resections have become more standardised, their indication in paediatric surgical oncology remains sporadic and controversial[3,4]. In addition to defining the indications for these extended resections, there is also lack of consensus regarding the technical aspects of these resections and reconstructions. Different strategies of vascular reconstruction are described in adult oncology, but few reports have been published in this regard in the paediatric population.[1,5] This paper describes our institutional experience with the surgical management of malignancies complicated by vascular invasion, including our techniques of resection and reconstruction, and discuss the current available literature.

Methods

After receiving approval from our Ethics Committee (HREC M220202) we performed a retrospective study from 1 January 2018 to 31 May 2022 including all patients younger than 18 years of age who were operated on for solid tumours with major abdominal vascular involvement which were selected for a concomitant vascular surgical procedure. We included those cases requiring resection of more than 50% of the circumference of a major abdominal vessel. Standardised procedures such as cavotomy for tumour thrombus removal or partial resection of less than 50% of a vessel wall were not considered for this study.

Demographic data, clinical and surgical information were retrospectively collected and reviewed. More specifically, age at surgery, type of tumour, pre-operative radiological findings and staging, location and side of the mass and vessels involved were collected.

Pre-operative assessment and management was directed according to tumour type and the respective institutional protocol. Surgical reports were retrospectively reviewed to collect intraoperative information, specifically the approach to resection of the primary disease, the vascular resection performed, including whether a reconstruction was performed and what type. Intra-operative complications were also noted. Histology reports of the resected specimen were reviewed to confirm the histological diagnosis of the primary tumour, and to assess the extent of vascular involvement. Post-operative management was evaluated with a specific focus on complications. All morbidity and mortality were reported, and long-term survival was recorded. Finally, we reviewed the available English literature specifically focusing on surgical management of vascular involvement in paediatric oncology.

Results

Seven patients met the criteria for this study, 4 male and 3 female. Average age was 8.25 years (range: 9-months – 14-years). Two patients presented with bilateral paraganglioma, 2 with pancreatic tumours, 2 with nephroblastoma, and 1 with hepatoblastoma (HBL). Patient characteristics are summarised in Table 1. Eight vascular procedures were performed: the 2 patients with Pancreatic tumours (1 pancreatoblastoma and 1 solid pseudopapillary tumour of the pancreas [SPN]) had portal vein (PV) resection and reconstruction with primary end-to-end anastomosis as part of a Whipple procedure. The remaining 5 patients had their procedures performed on the Inferior Vena Cava (IVC). Surgical information is summarised in Table 1.

Three patients had cavectomy with no reconstruction, 2 for nephroblastoma and 1 for HBL. The first Wilms Tumour (WT) was a 3-year-old male diagnosed with non-metastatic right-sided Wilms tumour with a tumour thrombus extending into the infrahepatic IVC. Neo-adjuvant chemotherapy was initiated according to the SIOP Umbrella protocol. However, the family refused surgery and absconded. The patient represented 6 months later with hypovolemic shock due to an intracapsular bleeding of a massive tumour requiring an emergency laparotomy. The IVC was not visible on the pre-operative CT scan, whilst the azygos vein was dilated. Intra-operatively, the infrahepatic supra renal IVC was not separable from the tumour which had infiltrated the IVC wall. Right nephrectomy with en-bloc cavectomy without reconstruction was performed. Although vascular margins of resection were free, the patient was staged as stage III for the pre-operative rupture. No post-operative complication was noted. However, the patient did not undergo radiation therapy and represented with extensive lung metastases, and he was referred for palliative care.

The 2nd patient with WT was a 7-year-old female previously treated for a stage 3 left sided tumour. At initial presentation, she had a tumour thrombus extending up the IVC into the heart, which regressed completely after neo-adjuvant chemotherapy. The patient subsequently underwent a standard left nephrectomy. Due to non-compliance, adjuvant chemo- and radiotherapy were not completed, and the patient represented 2 years later with a large relapse in the infrahepatic IVC. With extensive disease, and no demonstrable flow, she underwent an elective cavectomy without reconstruction. Histology confirmed positive resection margins, although of little relevance in this case, since second line chemotherapy and radiotherapy in recurrent disease are mandatory. The patient completed post-operative adjuvant treatment and is now disease free.

The last cavectomy was performed on a 9-month-old female who presented with a PRETEXT3VP HBL. After neo-adjuvant chemotherapy (NACT), she was reassessed as POSTTEXT3V due to IVC infiltration. She was presented to the transplant unit which assessed her as an unsuitable candidate for transplant, this based on the presence of persistent extrahepatic disease. An extended right hepatectomy with an En-bloc cavectomy was performed. No intra or post operative complications were noted. Whilst the vascular resection margin was tumour-free, microscopically residual disease was reported by the pathologist, this in the form of necrotic tumour cells present at the parenchymal resection margin. The patient underwent adjuvant chemotherapy and there has been no evidence of recurrence on serial follow-up CT scans.

Of the two patients who presented with bilateral paraganglioma infiltrating the IVC, one underwent bilateral resection of the mass together with a partial caval resection with primary reconstruction, as well as a partial resection and primary repair of the right renal artery. Complete resection was achieved and no complications occurred. The second patient with a paraganglioma underwent resection of the right sided tumour together with an infrarenal cavectomy and prosthetic graft (PTFE) interposition. This patient was complicated by an anastomotic bleed which required a relook laparotomy at 48 hours, as well as by graft infection 1-week post-surgery. At this procedure, the graft was clearly infected, completely thrombosed, and was removed, leaving the IVC interrupted. Post-operatively, the patient recovered and was subsequently discharged. Subsequently, a supraclavicular nodal metastasis with active uptake on MIBG was detected. The family refused surgery of the contralateral lesion, until he presented with severe haematemesis requiring emergency surgery and resection of the residual mass with a sleeve of duodenum as well as the kidney. The patient was lost to follow up thereafter.

The last 2 patients underwent Whipple procedures with concomitant resection and reconstruction of the Portal Vein for pancreatic head tumours: one Pancreatoblastoma and one SPN. The former underwent resection after 4 cycles of NACT for an unresponsive mass encasing the PV and coeliac trunk while the latter had a primary resection. In both cases, the Celiac Axis and Superior Mesenteric Artery were dissected off the tumour, while the PV was completely encased by the mass. In both cases, PV resection with end-to-end anastomosis was performed. Histology demonstrated that resection margins were clear of tumour in both cases. Post-operative Doppler demonstrated excellent portal flow in both cases. Both patients are doing well on follow-up.

Discussion

Oncovascular surgery is a recently introduced term, defined by Ghosh at al as cancer resection with concurrent ligation or reconstruction of a major vascular structure.[6] Oncovascular surgery is performed after considering the balance between the prognosis of the particular tumour and the expected morbidity of the procedure.[7]

Oncovascular Surgery is classified into three categories: surgery for tumours of primary vascular origin, rescue surgery for complications encountered during tumour resection, or planned en-bloc vascular and tumour resection.[8] This paper focuses on the latter category. The most common applications of this strategy in adult surgery are pancreatic tumours encasing the portal vein or the hepatic artery, retroperitoneal sarcomas invading major vasculature, or limb salvage surgery for extremity sarcomas.[8] Although oncovascular procedures are performed internationally for a wide range of oncological diseases in adults, no uniformity exists regarding strategy, operative technique and follow-up.[9] Oncovascular surgery is even more rare in children, and is only reported anecdotally.

With respect to arterial resection and reconstruction, whilst it has become common practice in specialised centres treating adult tumours, it remains rarely performed in children[10,6]. Resection of major arteries mandates reconstruction. Primary anastomosis is the preferred option but it is rarely feasible. Autologous graft with the iliac or saphenous vein, or prosthetic graft are the other options described.[8] Prosthetic grafts are

usually not preferred due to high risk of infection and thrombosis. However, they are the prevailing substitute for abdominal reconstruction[10]. A successful PTFE grafting of the infrarenal aorta for a recurrent ganglioneuroma in a 11-year old has been described.[11] Other arterial reconstructions in children are reported as part of bigger series including adult patients but are not discussed separately.[12,13]

Various childhood tumours are prone to vascular involvement. Neuroblastoma typically grows around vessels and the traditional recommendation is vessel definition, vessel clearance, followed by tumour removal ensuring that the integrity of the vessel is left intact[14]. Whilst vascular resection for neuroblastoma has been described, it remains controversial.[5] In our institution we do not perform vascular resections for neuroblastoma as the current evidence on the benefit of a complete resection (R0) does not add to a survival benefit, therefore we believe that the risks of resective vascular procedures outweigh the benefits[15]. On the contrary, for other malignancies, complete surgical resection remains the curative treatment goal[16].

Involvement of major venous vasculature such as the Inferior Vena Cava is particularly challenging. The most common tumours associated with invasion of the Vena Cava in childhood are hepatic and retroperitoneal malignancies, whereas direct intravascular extension and tumour thrombi are commonly associate with renal tumours, especially WT.[5,17] Thrombus removal in WT rarely requires vascular resection[18]. However, in rare occasions the thrombus is not separable from the vein wall and en bloc resection may be needed to achieve complete tumour resection.[19] The peculiarities of WT presenting to our institution make this scenario not uncommon. The higher incidence of WT in the African population together with a large rural community with poor access to health care make late presentation of these tumours common. As in a number of our cases, it is not uncommon for patients to default chemotherapy or radiotherapy, or refuse surgery, further contributing to disease progression and complications.

The IVC may also be involved by other retroperitoneum tumours such as teratomas and paragangliomas.[20,21] Pheochromocytomas and Paragangliomas can encase the IVC, occasionally requiring aggressive resection: cavectomy for invasive paragangliomas has been described in both adults and children.[16,22,21] Retroperitoneal sarcomas may also involve the IVC and cavectomy for such tumours is also reported.[5]

The IVC may be involved in Hepatoblastomas as well[23]. Involvement of the IVC is not an absolute contraindication to tumour resection, and en-bloc tumour resection with the IVC is contemplated in the SIOPEL4 protocol[24]. In our institution, relatively reduced access to transplantation together with significant experience in hepatobiliary surgery have induced a preference for more extensive primary resections of HBL unless absolutely contraindicated. The case in our series is an example of the success of advanced resection for a borderline resectable HBL, similar to what is reported for adults patients.[23,25]

With respect to invasion of the main portal vein in HBL, this is generally considered an absolute indication for transplant. PV resection, however, may be necessary for other paediatric malignancies, mostly pancreatic in origin.[26] Pancreatoblastoma and SPN are the most common pancreatic malignancies in childhood and hence the main indications for portal vein resection in children.[27,28] Both of these tumours are rare, hence specific protocols are not available. Surgical resection is the mainstay of treatment for both of these tumours, so when involved, PV resection is the only option to achieve complete tumour resection[26,29]. Generally, in cases presenting with vascular involvement, neo-adjuvant chemotherapy is ideal in an attempt to shrink the tumour and avoid these extended surgical techniques[1]. Of our 2 cases, the 7-year-old patient was reported to be a Pancreatoblastoma on the diagnostic biopsy, 4 cycles of neoadjuvant NACT were administered but with poor response. The second older patient had imagining consistent with an SPN and upfront surgery was performed.

For more common tumours like WT and HBL, protocols are available and they dictate neoadjuvant NACT[24,30]. On the contrary, Paragangliomas have typically a poor response to NACT and therefore primary en-bloc resection of mass and vessels must be considered[31,32,33,21]

After resection of the involved vessel, a decision needs to be made regards simple ligation, or reconstruction.[34] This will depend on whether an artery or vein is involved, and what it supplies: most arteries and the portal vein will require reconstruction, whilst the IVC can be ligated with impunity. Primary end-to-end anastomosis is the preferred option when feasible, as it is associated with improved outcomes[35], which is consistent with the 2 cases in our series. When primary anastomosis is not feasible, vascular reconstruction has been described, this including patch angioplasty, or interposition grafting using either autologous vein or prosthetic material.[36,34] There is currently no published consensus about the optimal graft for PV repair, although prosthetic graft appears to have a higher incidence of thrombosis.[37] Iliac vein[38], pericardium[27], and recanalized umbilical vein[39] have been described as options. In our institution, the preferred graft for PV reconstruction is the Internal Jugular Vein, this based on our experience with shunts for portal hypertension and transplant surgery. However, we haven't required interposition grafts in our portal venous reconstructions to date.

Prosthetic grafting for IVC reconstruction have been seldomly described in the paediatric population[5]. Grimaldi et al reported a series of 4 patients who underwent PTFE replacement of the IVC, 3 of whom developed graft thrombosis, although asymptomatic.[5] The high rate of complications after PTFE replacement of the IVC has been reported in adult literature too.[40] In our series, we utilised this strategy in one case, with the patient complicating with bleeding, and subsequent infection and thrombosis.

The alternative, resection without reconstruction, is well reported.[40] In adults presenting with IVC occlusion and no chronic venous disease, ligation is well tolerated, and may be associated with less blood loss and pulmonary thromboembolism.[41] Ligation relies on collateral venous drainage in the retroperitoneum, and it should be noted that extensive local resections may compromise further development of this venous drainage. For this reason, where the IVC is partially obstructed, or when pre-existing collaterals are disrupted intra-operatively, reconstruction is preferred.[40] Children tolerate ligation of the IVC particularly well, due to their ability to rapidly develop venous collaterals.[42,19,43] This is particularly true in cases of progressive occlusion due to tumour ingrowth, which allows the venous return to be gradually diverted through collaterals, usually demonstrated by a dilated azygos vein on pre-operative CT scan.[42,19,43]

Grimaldi et al reported routine prosthetic grafting of the IVC as a bridge solution to allow better recovery and development of collaterals as the graft slowly thromboses.[5] In our opinion this can be considered, however, the high risk of complications, including graft infection, is a significant consideration in favour of avoiding IVC reconstruction.[40] In our institution, if cavectomy is necessary, a trial of intraoperative caval exclusion is performed and reconstruction is considered only if the patient shows signs of haemodynamic instability. Another potential advantage of cavectomy without reconstruction is the avoidance of long term thromboprophylaxis.[19]

In adults, elective procedures performed with the aim of achieving complete resection including vascular resections did not demonstrate additional morbidity when compared to those patients that underwent tumour resection alone[1], unless the former were performed as emergencies[44]. Achieving complete resection is the ultimate goal for which oncovascular surgery is usually contemplated. In our series, this was possible in 5 of 7 cases, whilst in 2 patients histology showed microscopically positive resection margins. The HBL patient had tumour present at the hepatectomy margin, with the resection line at the absolute limit of leaving adequate functional liver with appropriate vascular inflow and outflow to the remaining segments. The caval vascular margin of resection was clear. Since leaving microscopically positive margin is acceptable in the context of complex liver resections[45,46], particularly when liver transplantation is contraindicated, or less accessible, this strategy allowed our patient to be cured. In our WT patient, cavectomy allowed us at least to achieve gross tumour resection, followed by adjuvant chemo and radiotherapy as per recognised treatment guidelines.[30]

Conclusion

Oncovascular paediatric surgery can allow the surgical team to achieve complete tumour resection in some cases of locally advanced tumours, previously deemed non-resectable. This may decrease the intensity of treatment and treatment-related-toxicity and in some cases change the aim of treatment from palliative to curative. Portal vein resection and reconstruction, cavectomy with or without reconstruction, are valid options in appropriately selected paediatric cases, with little morbidity. Whilst at present not enough data are available proving any long-term benefit in children, we believe this series of cases may support oncovascular procedures in particular paediatric cases. As more evidence emerges, hopefully it will be possible to identify criteria for selecting the patients who may be appropriate candidates for paediatric oncovascular surgical procedures.

References

1. DiPerna CA. Concomitant Vascular Procedures for Malignancies With Vascular Invasion. *Arch Surg*. 2002;137(8):901. doi:10.1001/archsurg.137.8.901

2. Pedrazzoli S, Pasquali C, Sperti C. General Aspects of Surgical Treatment of Pancreatic Cancer. *Dig Surg*. 1999;16(4):265-275. doi:10.1159/000018735

3. Delpero JR, Sauvanet A. Vascular Resection for Pancreatic Cancer: 2019 French Recommendations Based on a Literature Review From 2008 to 6-2019. *Front Oncol*. 2020;10:40. doi:10.3389/fonc.2020.00040

4. Evans DB, Farnell MB, Lillemoe KD, Vollmer C, Strasberg SM, Schulick RD. Surgical treatment of resectable and borderline resectable pancreas cancer: expert consensus statement. *Ann Surg Oncol*. 2009;16(7):1736-1744. doi:10.1245/s10434-009-0416-6

5. Grimaldi C, Bertocchini A, Crocoli A, et al. Caval replacement strategy in pediatric retroperitoneal tumors encasing the vena cava: a single-center experience and review of literature. *J Pediatr Surg*. 2019;54(3):557-561. doi:10.1016/j.jpedsurg.2018.06.008

6. Ghosh J, Bhowmick A, Baguneid M. Oncovascular surgery. *Eur J Surg Oncol EJSO*. 2011;37(12):1017-1024. doi:10.1016/j.ejso.2011.08.131

7. Cananzi FCM, Ruspi L, Fiore M, Sicoli F, Quagliuolo V, Gronchi A. Major vascular resection in retroperitoneal sarcoma surgery. *Surgery*. 2021;170(3):848-856. doi:10.1016/j.surg.2021.02.052

8. Han A, Ahn S, Min SK. Oncovascular Surgery: Essential Roles of Vascular Surgeons in Cancer Surgery. *Vasc Spec Int*. 2019;35(2):60-69. doi:10.5758/vsi.2019.35.2.60

9. Baláž P, Vikatmaa P, Björck M, Venermo M, Mani K, Whitley A. Oncovascular Surgery: The Current Situation and Future Perspectives in Europe. *Eur J Vasc Endovasc Surg*. 2022;63(2):350-351. doi:10.1016/j.ejvs.2021.11.015

10. Krutman M, Nishinari K, Pignataro BS, et al. Twenty years of experience in vascular reconstructions associated with resection of malignant neoplasms in a single cancer center. *J Vasc Surg*. 2019;69(6):1880-1888. doi:10.1016/j.jvs.2018.08.196

11. Fueglistaler P, Gurke L, Stierli P, et al. Major Vascular Resection and Prosthetic Replacement for Retroperitoneal Tumors. *World J Surg*. 2006;30(7):1344-1349. doi:10.1007/s00268-005-0555-2

12. Carpenter SG, Stone WM, Bower TC, Fowl RJ, Money SR. Surgical Management of Tumors Invading the Aorta and Major Arterial Structures. *Ann Vasc Surg*. 2011;25(8):1026-1035. doi:10.1016/j.avsg.2011.05.005

13. Poultsides GA, Tran TB, Zambrano E, et al. Sarcoma Resection With and Without Vascular Reconstruction: A Matched Case-control Study. *Ann Surg*. 2015;262(4):632-640. doi:10.1097/SLA.0000000000001455

14. Kiely EM. Radical surgery for abdominal neuroblastoma. *Semin Surg Oncol*. 1993;9(6):489-492. doi:10.1002/ssu.2980090606

15. Ryan AL, Akinkuotu A, Pierro A, Morgenstern DA, Irwin MS. The Role of Surgery in High-risk Neuroblastoma. *J Pediatr Hematol Oncol*. 2020;42(1):1-7. doi:10.1097/MPH.0000000000001607

16. Orbach L, Nachmany I, Goykhman Y, et al. Surgical Approach to Abdominal Tumors Involving the Inferior Vena Cava. *Isr Med Assoc J IMAJ*. 2020;22(6):364-368.

17. Al Diab A, Hirmas N, Almousa A, et al. Inferior vena cava involvement in children with Wilms tumor. *Pediatr Surg Int*. 2017;33(5):569-573. doi:10.1007/s00383-016-4034-7

18. Cox S, Büyükünal C, Millar AJW. Surgery for the complex Wilms tumour. *Pediatr Surg Int*. 2020;36(2):113-127. doi:10.1007/s00383-019-04596-w

19. Ceccanti S, Büyükünal C, Emre Ş, Masselli G, Schiavetti A, Cozzi DA. Resection of Inferior Vena Cava Without Reconstruction for Intravascular Intrusion of Wilms Tumor. *Urology*. 2021;149:e29-e33. doi:10.1016/j.urology.2020.11.004

20. Jones V, Burns C. Operative Considerations in Pediatric Retroperitoneal Teratomas—A Review. *Eur J Pediatr Surg*. 2012;23(04):265-269. doi:10.1055/s-0032-1313338

21. Srirangalingam U, Gunganah K, Carpenter R, Bhattacharya S, Edmondson SJ, Drake WM. Phaeochromocytoma and Paraganglioma Excision Involving the Great Vessels. *EJVES Short Rep*. 2017;35:1-6. doi:10.1016/j.ejvssr.2017.02.002

22. Mannina EM, Xiong Z, Self R, Kandil E. Resection of a Catecholamine-Elaborating Retroperitoneal Paraganglioma Invading the Inferior Vena Cava. *Case Rep Surg*. 2014;2014:1-4. doi:10.1155/2014/837054

23. Hemming AW, Reed AI, Langham MR, Fujita S, Howard RJ. Combined Resection of the Liver and Inferior Vena Cava for Hepatic Malignancy. *Ann Surg*. 2004;239(5):712-721. doi:10.1097/01.sla.0000124387.87757.eb

24. SIOPEL4 EUDRACT Number: 2004-001828-20. Final version July 2004.

25. Hemming AW, Reed AI, Fujita S, Zendejas I, Howard RJ, Kim RD. Role for Extending Hepatic Resection Using an Aggressive Approach to Liver Surgery. *J Am Coll Surg*. 2008;206(5):870-875. doi:10.1016/j.jamcollsurg.2007.12.036

26. d'Ambrosio G, del Prete L, Grimaldi C, et al. Pancreaticoduodenectomy for malignancies in children. *J Pediatr Surg*. 2014;49(4):534-538. doi:10.1016/j.jpedsurg.2013.09.010

27. Yonekura T, Kosumi T, Hokim M, Hirooka S, Kitayama H, Kubota A. Aggressive surgical and chemotherapeutic treatment of advanced pancreatoblastoma associated with tumor thrombus in portal vein. *J Pediatr Surg*. 2006;41(3):596-598. doi:10.1016/j.jpedsurg.2005.11.071

28. Yu P, Cheng X, Du Y, et al. Solid Pseudopapillary Neoplasms of the Pancreas: a 19-Year Multicenter Experience in China. *J Gastrointest Surg*. 2015;19(8):1433-1440. doi:10.1007/s11605-015-2862-8

29. Wang KS, Albanese C, Dada F, Skarsgard ED. Papillary cystic neoplasm of the pancreas: A report of three pediatric cases and literature review. *J Pediatr Surg*. 1998;33(6):842-845. doi:10.1016/S0022-3468(98)90655-8

30. UMBRELLA PROTOCOL SIOP-RTSG 2016. EudraCT number: 2016-004180-39.

31. Hu H, Huang B, Zhao J, Yuan D, Yang Y, Xiong F. En Bloc Resection with Major Blood Vessel Reconstruction for Locally Invasive Retroperitoneal Paragangliomas: A 15-Year Experience with Literature Review. *World J Surg*. 2017;41(4):997-1004. doi:10.1007/s00268-016-3846-x

32. Lenders JWM, Duh QY, Eisenhofer G, et al. Pheochromocytoma and Paraganglioma: An Endocrine Society Clinical Practice Guideline. *J Clin Endocrinol Metab*. 2014;99(6):1915-1942. doi:10.1210/jc.2014-1498

33. Waguespack SG, Rich T, Grubbs E, et al. A Current Review of the Etiology, Diagnosis, and Treatment of Pediatric Pheochromocytoma and Paraganglioma. *J Clin Endocrinol Metab*. 2010;95(5):2023-2037. doi:10.1210/jc.2009-2830

34. Ruiz CS, Kalbaugh CA, Browder SE, et al. Operative strategies for inferior vena cava repair in oncologic surgery. *J Vasc Surg Venous Lymphat Disord*. 2020;8(3):396-404. doi:10.1016/j.jvsv.2019.09.012

35. Dua MM, Tran TB, Klausner J, et al. Pancreatectomy with vein reconstruction: technique matters. *HPB*. 2015;17(9):824-831. doi:10.1111/hpb.12463

36. Gaignard E, Bergeat D, Robin F, Corbière L, Rayar M, Meunier B. Inferior Vena Cava Leiomyosarcoma: What Method of Reconstruction for Which Type of Resection? *World J Surg*. 2020;44(10):3537-3544. doi:10.1007/s00268-020-05602-2

37. Labori KJ, Kleive D, Khan A, Farnes I, Fosby B, Line PD. Graft type for superior mesenteric and portal vein reconstruction in pancreatic surgery – A systematic review. *HPB*. 2021;23(4):483-494. doi:10.1016/j.hpb.2020.11.008

38. Kanai T, Miyazaki S, Akatsu T, Nakagawa M, Kameyama K. Long-term Follow-up after Pancreatoduodenectomy with Portal Vein Resection for a Huge Solid Pseudopapillary Neoplasm in an Adolescent Girl. *Keio J Med*. 2014;63(3):52-59. doi:10.2302/kjm.2014-0003-CR

39. Shinkai M, Mochizuki K, Kitagawa N, et al. Usefulness of a recanalized umbilical vein for vascular reconstruction in pediatric hepatic surgery. *Pediatr Surg Int*. 2016;32(6):553-558. doi:10.1007/s00383-016-3893-2

40. Wysocki AP, Hetherington R, Nicol D, Gibbs HH. Haemodynamic assessment following inferior vena cava resection without replacement. *ANZ J Surg*. 2004;74(8):667-670. doi:10.1111/j.1445-1433.2004.03127.x

41. Mathisen DJ, Javadpour N. En bloc resection of inferior vena cava in cytoreductive surgery for bulky retroperitoneal metastatic testicular cancer. *Urology*. 1980;16(1):51-54. doi:10.1016/0090-4295(80)90331-3

42. Jones VS, Shun A. Is the inferior vena cava dispensable? *Pediatr Surg Int*. 2007;23(9):885-888. doi:10.1007/s00383-007-1969-8

43. Ribeiro RC, Schettini ST, Abib S de CV, da Fonseca JHP, Cypriano M, da Silva NS. Cavectomy for the treatment of Wilms tumor with vascular extension. *J Urol*. 2006;176(1):279-283; discussion 283-284. doi:10.1016/S0022-5347(06)00561-1

44. Bianchi C, Ballard JL, Bergan JH, Killeen JD. Vascular reconstruction and major resection for malignancy. *Arch Surg Chic Ill 1960*. 1999;134(8):851-855. doi:10.1001/archsurg.134.8.851

45. Aronson DC, Weeda VB, Maibach R, et al. Microscopically positive resection margin after hepatoblastoma resection: what is the impact on prognosis? A Childhood Liver Tumours Strategy Group (SIOPEL) report. *Eur J Cancer*. 2019;106:126-132. doi:10.1016/j.ejca.2018.10.013

46. Younes A, Elgendy A, Fadel S, et al. Surgical Resection of Hepatoblastoma: Factors Affecting Local Recurrence. *Eur J Pediatr Surg*. 2021;31(05):432-438. doi:10.1055/s-0040-1717087

Table 1

TABLE 1

Patient n	Sex	Age	Diagnosis	Vessel invasion	NACT	Procedure	Radical resection	Complications	Outcome
1	F	14 yr	SPN of pancreas	Portal vein, hepatic artery	No	Whipple pr + PV resection and anastomosis; hepatic artery dissected from toumor	R0	No	Alive at 4 yrs FU
2	M	7 yr	Pancreatoblastoma	Portal vein, coeliac trunk	4 cycles	Whipple pr + PV resection and anastomosis; coeliac trunk dissected from toumor	R0	No	Alive at 1 yr FU
3	M	11 yr	Bilateral Paraganglioma	IVC, right renal artery, coeliac trunk	No	Resection, dissection of arteries, partial cavectomy with primary repair	R0	No	Alive at 4 yrs FU
4	M	14 yr	Bilateral Paraganglioma	IVC	No	Tumour resection, cavectomy with PTFE grafting	R0	Haemodynamic shock for anastomotic leak; graft infection and thrombosis	Alive at 3 yrs FU
5	M	4 yr	Right Wilms tumour	IVC	Defaulted	En-bloc right nephrectomy + suprarenal cavectomy no reconstruction	R0	No	Alive at 3 yr FU
6	F	7 yr	Relapse of left Wilms tumour	Tumour relapse in suprarenal IVC	Yes	Cavectomy with no reconstruction	R1	No	Alive at 6 months FU
7	F	9 month	Hepatoblastoma	Retrohepatic IVC	6 cycles	En.-bloc extended right hepatectomy and cavectomy with no reconstruction	R1	No	Alive at 1 yr FU