

## Aortic Replacement for Retroperitoneal Tumors in Children

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Received: 20 Oct 2021

Accepted: 16 Nov 2021

Published: 20 Nov 2021

J Short Name: COO

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### Keywords:

Retroperitoneal neoplasms; Ganglioneuroma; Paraganglioma; Aorta; Abdominal surgery

### Citation:

Rodrigo Chaves Ribeiro, Aortic Replacement for Retroperitoneal Tumors in Children. Clin Onco. 2021; 5(6): 1-5

## 1. Abstract

Retroperitoneal tumors may involve the abdominal aorta. Sometimes, these tumors are unresponsive to adjuvant therapy. The total resection is a challenge mainly for vascular reconstruction in children. Then, we report three cases of aortic replacement in children with retroperitoneal tumors. Case 1: an eight-year-old girl presenting with a ganglioneuroma involving the aorta. The tumor was resected with an abdominal aorta segment, which was replaced by an aorto-aortic prosthesis. Case 2: a 13-year-old boy was presented with a paraganglioma involving the aortic bifurcation, requiring resection and replacement by aortoiliac bypass. Case 3: an 11-year-old girl with abdominal neurofibromatosis. During the resection, the aorta and left iliac artery were replaced by an aortoiliac graft. Resection of a segment of the aorta and revascularization using a prosthesis is feasible in retroperitoneal tumors in children, but the long-term results are unknown.

## 2. Introduction

Retroperitoneal tumors in children may involve or surround the abdominal aorta and its main branches. Such vascular involvement leads to challenging surgical procedures and may be considered unresectable. When large tumors involve major vascular trunks and do not respond to chemotherapy, resection of the tumor with

replacement of the aorta may be an alternative. Replacement of the abdominal aorta in children is a complex procedure that is rarely performed. In children, synthetic grafts are limited by a concern for late infection and lack of potential growth. There are few reports on this in the literature. We report three cases of children with retroperitoneal tumors, in which replacement of the aorta was needed in order to resect the tumor.

## 3. Patients and Methods

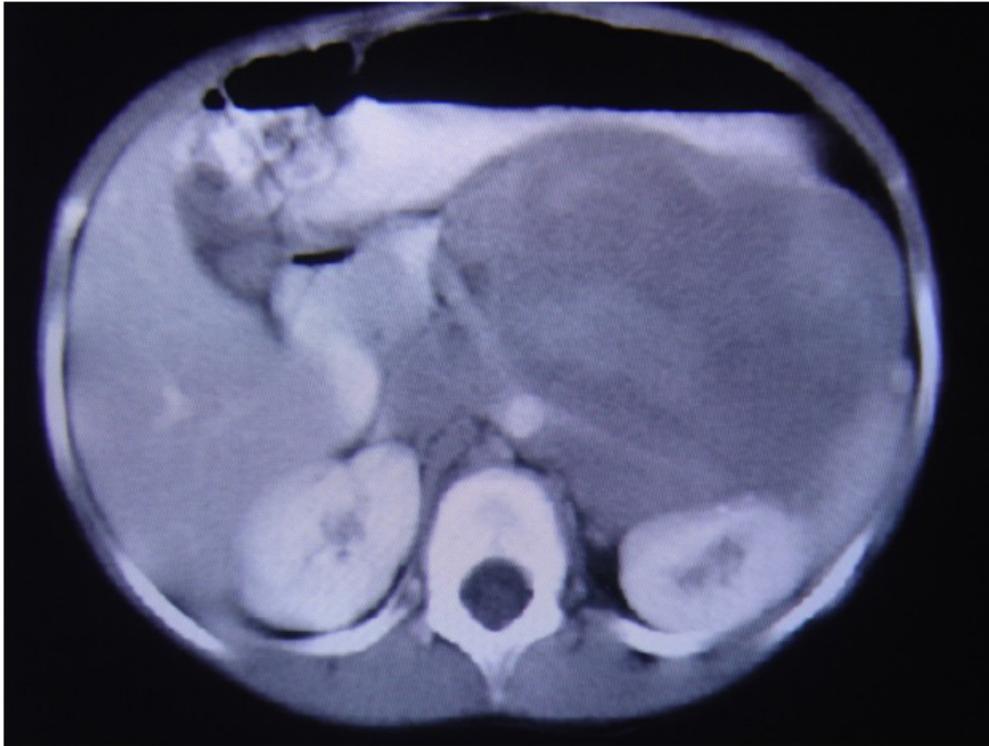
Cases came from two reference institutions for pediatric oncology, pediatric surgical oncology, and vascular surgery. Three cases in which tumor resection was performed with replacement of the aorta or its branches were found, who underwent three procedures.

## 4. Case 1

An eight-year-old girl, presented with abdominal mass for 10 months and deficit of growth for two years. Ultrasound (US) and computed tomography (CT) revealed a mass involving the aorta, celiac trunk, mesenteric artery, and left renal hilum (Figure 1). Initial biopsy revealed a ganglioneuroma. Due to tumor dimension and suspecting that there might be concomitant malignant parts within the tumor that might not have been not sampled at the initial biopsy, a partial resection (debulking) procedure was indicated. During this procedure, the aorta and superior mesen-

teric artery were injured. The tumor was resected with a segment of the abdominal aorta and the left kidney. The mesenteric artery was reimplanted in the aorta. The aorta was clamped in its suprarenal segment for 40 minutes. The vascular reconstruction was done with an aorto-aortic Dacron prosthesis. Renal failure did not develop postoperatively, but the patient presented enterorrhagia until the second postoperative day, which was attributed to an ischemic

lesion of the intestinal mucosa. She also had chronic diarrhea until six months after surgery, and platelet antiaggregant (acetylsalicylic acid) was used for three months after the operation. The patient remains free of recurrences, and there are no vascular and gastrointestinal symptoms, nor vascular deficits or limb impairment 40 months after surgery. Doppler controls are normal.



**Figure 1.** Abdominal tomography of huge ganglioneuroma. The tumor surrounding the aorta and the superior mesenteric artery.

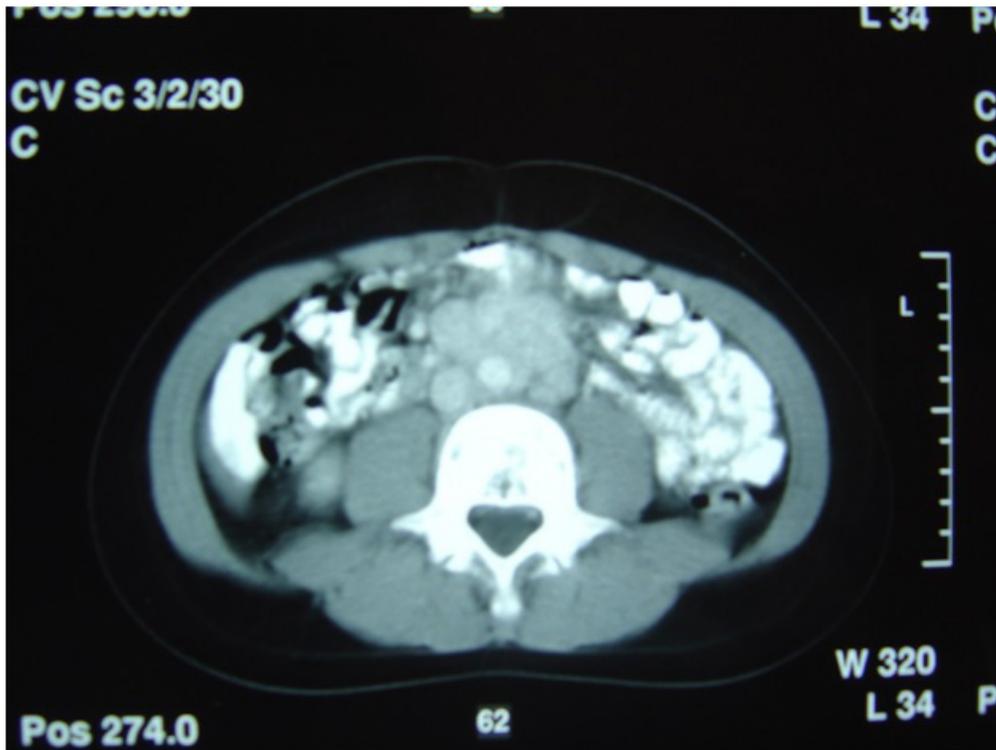
## 5. Case 2

A 13-year-old boy was presenting an episodic crisis of headache, hypertension, and sweating. At admission, an abdominal mass was palpated in the mesogastrium. US, CT, and nuclear magnetic resonance (NMR) imaging examinations revealed a mass involving the bifurcation of the aorta (Figure 2). The urinary metabolites of catecholamines were elevated. Initial biopsy revealed a paraganglioma. Preoperative preparation using an alpha-adrenergic blocker was administered for ten days. During the operation, the mass surrounded the aorta, and an aortic resection was necessary to obtain complete excision. The aorta was clamped in its infrarenal portion, and reconstruction was done using an aortoiliac Dacron graft (Figure 3). The patient had thrombosis of the left iliac vein postoperatively and was kept on anticoagulants for 30 days after the operation. One year after the initial surgery, he presented a recurrence of the episodic symptoms of headache, hypertension,

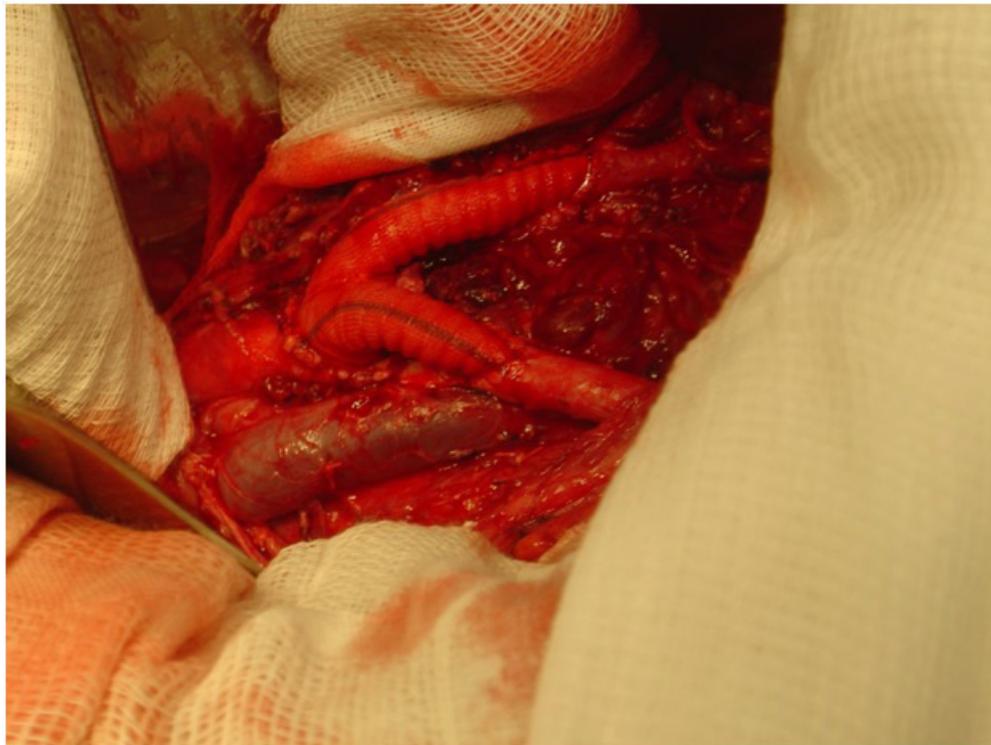
and sweating. Abdominal CT revealed recurrence at the tumor site. He underwent reoperation with complete resection of the tumor, which was posterior to the aortic bypass. It was not possible to separate it from the left iliac vein, and the resection included a segment of the referred vein. The patient is free of symptoms, and there are no sequelae, vascular deficit, or limb impairment 30 months after the second operation.

## 6. Case 3

An 11-year-old girl with type 1 neurofibromatosis presented with an abdominal mass in the left iliac fossa. The biopsy showed a Triton tumor. Therefore, a mass resection procedure was indicated, during which the aorta and left iliac artery were injured and were replaced with an aortoiliac polytetrafluoroethylene graft. The iliac vein was ligated. Pathology revealed a peripheral nerve sheath tumor with clusters of rhabdomyosarcoma. The patient died four months after surgery, due to disease progression.



**Figure 2.** Abdominal tomography of parangioma. The tumor surrounding the aortic byfurcation.



**Figure 3.** Aortobiliac Dacron Graft.

## 7. Discussion

Vascular surgery in children is challenged by the small vessel size and compensation for future growth in diameter and length. Synthetic grafts provide good long-term durability, but there are concerns about late infections and pressure gradients.

Replacement of the abdominal aorta in children has already been described in certain vascular diseases, such as congenital aneurysms of the abdominal aorta [1, 2], pseudoaneurysm due to hydatid cyst [3], and middle aortic syndrome [4]. Despite the initial success in the reported cases of replacement of the aorta in children, there are few descriptions of the late outcome of these patients. It is likely that prosthesis replacement will be needed if the child's growth causes a significant impact on the pressure gradient in the graft. However, the development of collateral circulation may minimize these effects.

Barral et al. [5] described eight cases of replacement of the aorta during childhood because of a middle aortic syndrome, hypoplasia aortic, and aortic aneurysm, with long-term patient follow-up [5]. Only three patients required graft revision after lengthy follow-up. Meyers described the use of a cryopreserved and decellularized homologous graft in two children [6]. Over the course of the follow-up, there was a slight increase in the diameter of this graft. A long-term follow-up would be needed in order to evaluate this type of graft more precisely.

We found few reports on aorta replacement for treating abdominal tumors in children in the literature. Meyers et al described aorta replacement with resection of neuroblastoma of the organ of Zuckerkandl in a four-year-old boy [6], like in our second case, which was a paraganglioma of the Zuckerkandl organ. Ito et al described resection of the aorta and vena cava in a child with retroperitoneal rhabdomyosarcoma Paran et al. [7] described five children who sustained aorta injuries during resection of retroperitoneal neuroblastomas [8].

Retroperitoneal tumors with involved major vessels can be resected when vascular reconstruction is possible. Even for adults, the literature is poor, and there are just cases series [9, 10, 11]. However, in these cases, the surgery could be curative. This report is crucial to demonstrate that it is possible to do this kind of surgery in children.

There is a series of fifteen adults cases of resection of paraganglioma with major vessels, and there was aorta involved. The 5-year survival was 74%. However, there is no survival in a comparative group (patients that refuse the surgery) [12]. There was a description of two-stage surgery; the first is the arterial bypass followed by the resection surgery [10]. Homsy et al. [10] in a recent study, described seven aortic resections in seventh casuist patients with abdominal sarcoma major vessel involvement. They concluded that oncovascular surgery enables radical resection required for reasonable local control of retroperitoneal sarcomas and is associated

with an acceptable level of complications peri-operatively and during follow-up [13]. We believe this conclusion can be transposed to pediatric patients.

Another similar discussion is about cava resection; besides being a great vessel, not necessary to do reconstruction because of collateral formation. But how to aorta resection to tumor involving the literature is based on case series [14,15]

Our first two patients underwent resection of the aorta due to ganglioneuroma and paraganglioma, respectively. Follow-up was of 40 months (case 1) and 30 months (case 2). Both patients present good outcomes, but the possibility that a pressure gradient may develop remains, with the possible need for a graft change procedure. The third patient in our series had an initial diagnosis of neurofibromatosis, but malignant transformation occurred, and she died due to disease progression.

We believe that if there is a localized disease surrounding the large vessels, without distant metastasis. When no more adjuvant therapy can be administered, tumor resection with resection of a large abdominal vessel can be attempted. The possibilities for vascular reconstruction need to be analyzed and planned, and one has to bear in mind that the surgical risks in this type of procedure are high.

## 8. Conclusion

Resection of a segment of the aorta and revascularization using a prosthesis is feasible in retroperitoneal tumors in children, but the long-term results are unknown. A later analysis is necessary in order to conclude the benefits of these procedures.

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