Journal of
Medical Case Reports

CASE REPORT

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Early experience with renal autotransplant for renal artery stenosis in a 6-year-old patient with neurofibromatosis type 1: a case report

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Abstract

Background Renal artery stenosis due to neurofibromatosis type 1 is a known important source of secondary renovascular hypertension in pediatric patients. There are no guidelines on the management of renal artery stenosis in children, and the utility of stents and bypass grafting is limited given small patient size. Renal autotransplant to treat renal artery stenosis in a small pediatric patient may be a viable alternative for treatment and spare the need for nephrectomy.

Case presentation In this article, we present a case of renal autotransplant in a 6-year-old, 15.8 kg Nepali patient with neurofibromatosis type 1 with refractory hypertension and high-grade stenosis of the proximal right main renal artery. The patient underwent balloon angioplasty, which failed to dilate the stenosis. He later developed hypertensive urgency and required admission to the pediatric intensive care unit. The patient was not a candidate for repeat angioplasty given the length of the stenotic segment and its tortuosity. Blood pressure was unable to be controlled on multiple antihypertensive agents and the patient eventually developed hypertensive urgency. Therefore, a renal autotransplant of the right kidney was performed after multidisciplinary evaluation. The right renal artery ostium had significant hypertrophied intima involving 50% of its circumference. The kidney was procured in the same fashion as a living kidney donor nephrectomy. The kidney was mobilized *in situ*, and heparin was administered. The renal artery and renal vein were divided with surgical staplers. The kidney was removed from the patient and moved to the back table. On the back table, the organ was flushed with cold organ preservation solution and vessels inspected. The diseased portion of the right renal artery was resected to the location of no gross intimal thickening. It was judged that there was adequate length of the healthy artery remaining to allow safe reimplantation. The renal artery and vein were reimplanted to the abdominal aorta and inferior vena cava, respectively. The patient tolerated the surgery well, and 2 years postoperatively, he only requires one antihypertensive medication.

Conclusion Nephrectomy may be favored over renal autotransplant in small pediatric patients due to technical difficulties associated with autotransplant. We demonstrate significant clinical improvement in blood pressure control in a 15.8 kg, 6-year-old pediatric patient after renal autotransplant.

Keywords Neurofibromatosis, Renal artery stenosis, Renovascular hypertension, Renal autotransplant, Pediatric, Case report

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Background

Neurofibromatosis type 1 (von Recklinghausen disease; NF1) is an autosomal dominant, multisystem, neurogenetic disorder. It has characteristic clinical features such as café-au-lait spots, skinfold freckling, cutaneous neurofibromas, and Lisch nodules. NF1 can also lead to vasculopathies, such as renal artery stenosis (RAS) and abdominal aorta coarctation, which may manifest as renovascular hypertension (HTN). RAS is the most common site for symptomatic vasculopathy and occurs in approximately 2% of patients with NF1 [1, 2]. HTN secondary to RAS is initially treated with medications and endovascular intervention. However, patients with NF1 appear to have a lower success rate for renal artery angioplasty than non-NF1 patients [3]. Surgical options for RAS include nephrectomy and renal autotransplant, which may be less favored due to technical factors in a small pediatric patient. In this case report, we describe successful treatment of RAS in a small pediatric patient with NF1 using renal autotransplant.

Case presentation

A 6-year-old Nepali patient presented to the Nephrology Clinic at our institution with HTN noted at multiple office visits. He was known to have NF1, with multiple café-au-lait spots, an optic glioma, and a low-grade brain glioma. Blood pressure (BP) readings at their office appointments prior to presentation ranged from 139–154 mmHg to 92–105 mmHg. His family history was significant for maternal HTN, diagnosed at age 36 years old, and a sibling with NF1. A home BP monitor and cuff were issued for home readings, and a laboratory and imaging evaluation were undertaken. Electrolytes, renal function, renin, aldosterone, and metanephrines were all within normal limits. Home systolic BPs continued to be between 140 mmHg and 150 mmHg, and 1 week following initial nephrology appointment, computed tomography (CT) angiography of the renal vessels (Fig. 1) was carried out, revealing high-grade stenosis of the proximal right main renal artery and normal renal parenchyma and size. Our patient was then started on amlodipine 5 mg daily and home BP decreased to 110–120/70–90 mmHg. Our patient underwent elective bilateral renal angiography (Fig. 2) with dilation and renal artery balloon angioplasty with interventional radiology (IR). A 4.5×20 mm balloon was used in the right main renal artery for dilation. No stent was placed. It was noted during the procedure that there was incomplete dilation of the stenotic segment despite inflation to maximum rated balloon pressure, presumably due to the fibrotic nature of the stricture, but the IR team achieved overall improved appearance of the stenosis. At 2-week follow-up visit, 5 mg lisinopril daily was started for average home BPs greater than 120/80 mmHg. The patient presented to our Emergency Department with preorbital cellulitis. BP was elevated during this visit, and after a brief attempt to bring BP down to an ambulatory setting, our patient was admitted to the pediatric intensive care unit (PICU). Repeat CT angiography demonstrated very high-grade right main artery stenosis measuring 2 mm, with post-stenotic dilatation. IR deemed our patient to not be a candidate for a repeat attempt at balloon angioplasty. The abdominal transplant surgery team reviewed surgical

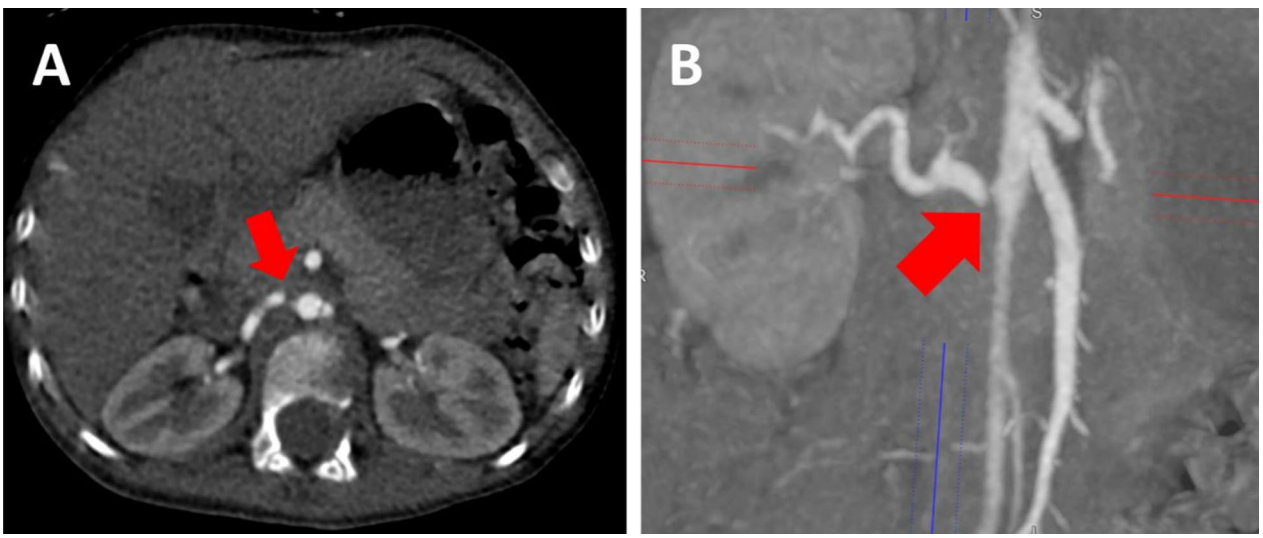

Fig. 1 Representative (**A**) sagittal and (**B**) coronal image from computed tomography angiography of the abdomen. The red arrow demonstrates high-grade stenosis in the proximal right renal artery

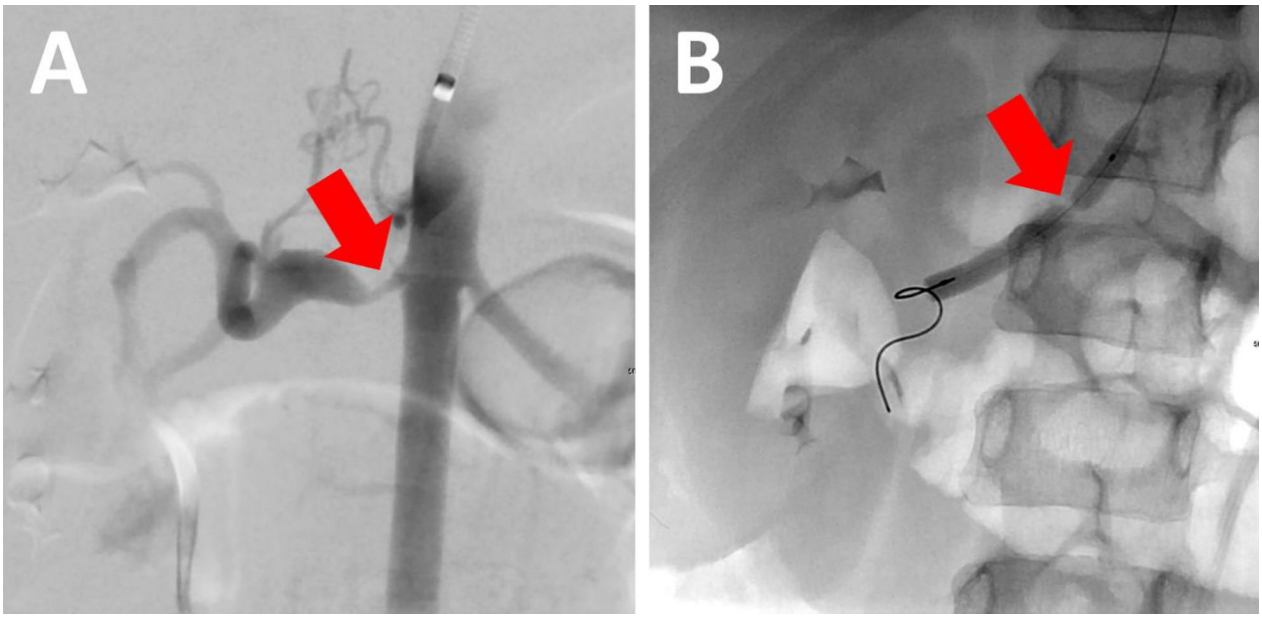

Fig. 2 Renal arteriogram. Conventional subtracted angiogram image (**A**) demonstrating stenosis (red arrows) at the origin of the right renal artery. Subsequent balloon dilation of the lesion (**B**) with a 4.5 mm diameter balloon shows incomplete dilation of the lesion despite application of maximum rated balloon pressure

options. A nuclear medicine renal scintigraphy scan with Lasix showed that the patient had split renal function of 55% on the left and 45% on the right. Multidisciplinary review of the case determined a renal autotransplant would potentially offer the maximum benefit, avoiding the need for a nephrectomy. Renal autotransplant was undertaken via a midline laparotomy incision, and a Cattell–Braasch maneuver was performed to expose the inferior vena cava (IVC) and right kidney. The right renal artery was identified and radiological findings of arterial stenosis was confirmed. The kidney was judged to be re-implantable, with an acceptable length of healthy artery present. The right kidney, including its vessels and ureter, was mobilized. Prior to ligating the right renal artery and vein, the patient was anti-coagulated with 800 units of heparin. The ureter was divided. We then divided the renal artery, and then the renal vein with surgical staplers. The right kidney was removed and placed on the back table, where it was flushed with a preservation solution via the renal vein at 4 °C. The renal artery ostium had significantly hypertrophied intima involving 50% of its circumference. The renal artery was resected to a level with no gross intimal thickening. The renal artery was flushed with 500 mL of preservation solution. The organ quickly achieved a very uniform pale color, indicating a high-quality flush.

Before implantation of the right kidney, the patient received 500 more units of heparin. The distal IVC was clamped and a running end-to- side anastomosis between the right renal vein and anterior side of the IVC was created (Fig. 3). Subsequently, the distal aorta was clamped, and a running end-to-side anastomosis was constructed for the right renal artery and distal aorta. The clamps were removed, and the organ reperfused without any complications. The right kidney immediately turned pink with even turgor and no mottled areas. The ureteroneocystostomy was constructed and a 3.8 French double J-ureteral stent was placed in the ureter and bladder before completion of the anastomosis. The abdominal fascia and skin were closed.

Postoperatively, the patient was placed on a nicardipine drip for BP control with a goal systolic BP of 110–130 mmHg (Fig. 4). On postoperative day 3, the patient was weaned off nicardipine drip, transferred to the ward, and transitioned to oral labetalol 100 mg twice daily and amlodipine 5 mg daily. These agents were weaned throughout hospital stay due to improved BP control. The patient was also started on a prophylactic heparin drip (10 units/kg/hour) and switched to aspirin 81 mg daily on postoperative day 4. At discharge (postoperative day 13), patient was requiring only amlodipine 2.5 mg daily and aspirin 81 mg daily. They also underwent serial ultrasound imaging of the right kidney after the renal autotransplant, which showed normal right kidney and associated vasculature.

After discharge, the patient was followed up with nephrology and abdominal transplant surgery team. He still had stage 1 HTN more than 1 month after surgery,

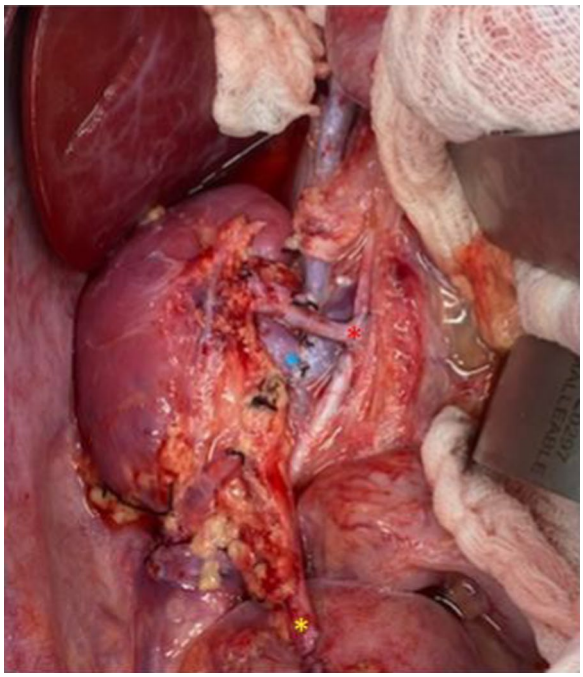

Fig. 3 Intraoperative photo of autotransplanted kidney after reperfusion. Intraoperative image of the right kidney re-transplanted after *ex vivo* excision of stenotic segment of the proximal right renal artery. The blue asterisk highlights the right renal vein re-anastomosed to the inferior vena cava. The red asterisk indicates the right renal artery re-anastomosed to the aorta. The yellow asterisk demonstrates the ureteroneocystostomy

therefore amlodipine dose was increased to 3 mg twice daily; 24 months after right renal autotransplant, home systolic BP measurements ranged from 90 mmHg to 100 mmHg.

Discussion and conclusion

The incidence of NF1 is approximately 1 in 3000 births [4]. Patients with NF1 may develop cardiac and vascular disease. Cardiovascular disease is a frequent cause of premature death in this patient population [5]. The second leading cause of death in patients with NF1 are vasculopathies, including aneurysms, arterial stenoses, and arteriovenous malformations [6]. The overall incidence of HTN in patients with NF1 is approximately 16% [7]. These patients typically develop renovascular HTN secondary to RAS, which represents a clinical challenge due to the ostial localization and presence of tough fibrotic tissue, often with long stenotic segments [8].

Early diagnosis and expedient management of renovascular HTN are imperative in children with NF1. If patients do not achieve BP control after medical optimization and endovascular intervention, then they may undergo nephrectomy, renal autotransplant, or bypass. However, there are no current guidelines for surgical management of RAS in pediatric patients with NF1. Nephrectomy may not be the best option in such patients because arterial disease can recur in the remaining kidney. However, it may be favored over more technically challenging surgeries, such as renal autotransplant, particularly in small pediatric patients. A retrospective review by Eliason *et al.* (2016) found that children who had a failed endovascular intervention were

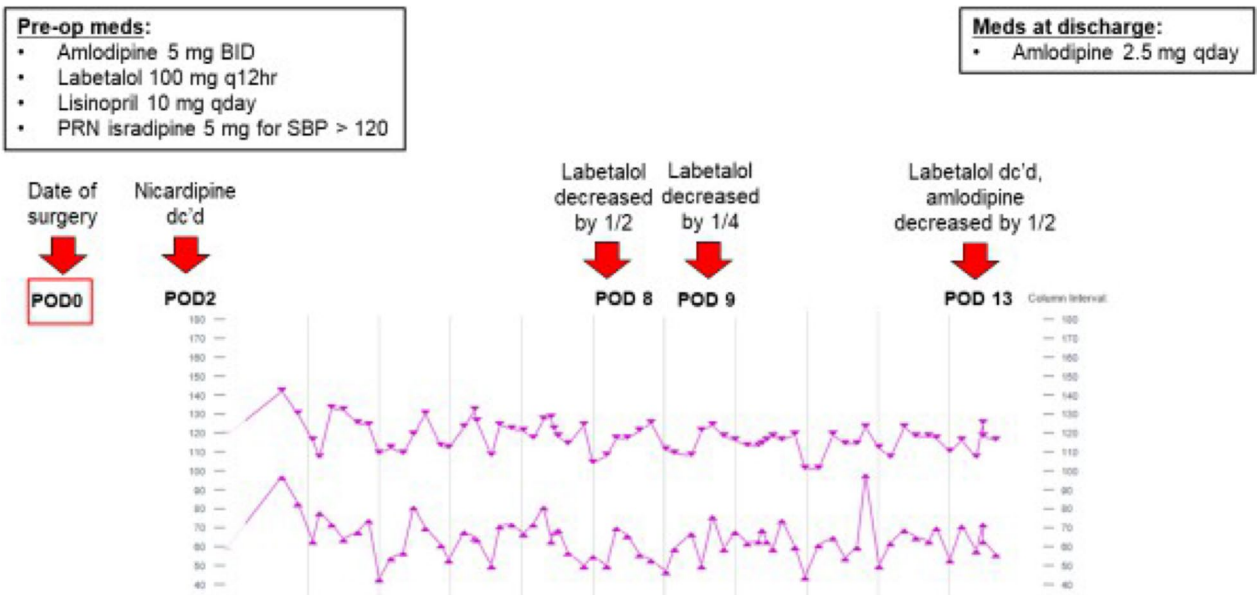

Fig. 4 Postoperative hypertension management and de-escalation of therapy. Postoperative timeline of blood pressure measurements and de-escalation of anti-hypertensives until day of discharge on postoperative day 13

significantly more likely to undergo a nephrectomy if they were younger than 10 years old [9]. A 40% nephrectomy rate in children younger than 10 years old was reported. In the same study, they found no benefit to undergoing a nephrectomy compared with arterial reconstructive surgery in patients with unilateral renal artery disease [9]. Another retrospective review examined ten pediatric patients with renovascular HTN and isolated RAS and found that nine of them underwent renal autotransplant, with the youngest being 7 years old [10]. One 5.5-year-old patient with isolated RAS underwent nephrectomy. Nonetheless, there has been a trend toward surgical revascularization over nephrectomy [11].

If there is no irreparable kidney or vascular disease, we posit that renal autotransplant should be considered in pediatric patients despite their age and size because this procedure may offer more optimal long-term outcomes compared with nephrectomy. Despite the high-grade stenosis in the right renal artery, our patient's right kidney had normal parenchyma and contributed 45% to the overall renal function, determined by preoperative ^{99m}Tc-MAG3 renography. Renal autotransplant also offers the advantage of preserving the patient's native nephron mass, which is desirable in a growing child. In addition, a retrospective case review demonstrated improved normalization of BP with angioplasty after autotransplant in pediatric patients with NF1 [12]. However, there are risks associated with this procedure. For instance, similar to allogenic renal transplant, prolonged ischemia time is associated with delayed renal function [13]. Therefore, meticulous perioperative and intraoperative planning and communication to limit cold and warm ischemia time is essential to optimal outcomes. In our case, warm ischemia time was minimized by procuring the kidney in a manner similar to living donor nephrectomy. We were able to examine the kidney after flush with cold preservative solution, and precisely resect the diseased segment of renal artery. Our cold ischemia and warm ischemia times were similar to those expected in living donor renal transplant, and our patient had immediate function of his autotransplanted kidney. The timing for renal autotransplant remains controversial. Some authors advocate for renal revascularization in children older than 3 years of age to avoid reoperation [14, 15]. A retrospective review with 13 pediatric patients with renal vascular disease found that the average age was 8.5 years (range 4–12 years) at consultation for HTN and 9.12 years (range 6–13 years) at time of renal autotransplantation [8]. The study did not discuss the reason for the gap in time between consultation and autotransplant. Other centers, reported by Jordan *et al.* [16], urge immediate repair of the renal artery disorder in pediatric patients to avoid progressive renal disease due to suboptimal HTN control or ischemic atrophy secondary to renal artery obstruction [16]. We believe that multidisciplinary assessment is crucial. Treatment should be individualized to the patient while considering the patient's age, severity of renovascular HTN, and specific vasculopathy [15]. Future investigations focused on the youngest age at which renal autotransplant offers success in experienced hands are warranted.

In this case report, we showed significant improvement in BP control in a 15.8 kg, 6-year-old patient who underwent renal autotransplant for high-grade renal artery stenosis. We recommend children with RAS to be referred to a multidisciplinary team, including pediatric transplant surgeons, for surgical evaluation to potentially avoid unnecessary nephrectomy.

Abbreviations

RAS	Renal artery stenosis
NF1	Neurofibromatosis type 1
HTN	Hypertension
BP	Blood pressure
IR	Interventional radiology

Acknowledgements
Not applicable.

Author contributions
All authors have contributed to the composition of this manuscript and its revisions. All authors have participated in terms of planning, design, writing and editing, and have approved the final manuscript.

Funding
Not applicable.

Availability of data and materials
The data presented here and/or used during the study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Written informed consent was obtained from the patient's parents for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors have no relevant financial interests and no conflicts of interest to disclose.

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Received: 12 August 2024 Accepted: 19 March 2025
Published online: 29 April 2025

References

1. Dubov T, Toledano-Alhadef H, Chernin G, Constantini S, Cleper R, Ben-Shachar S. High prevalence of elevated blood pressure among children with neurofibromatosis type 1. Pediatr Nephrol. 2016;31(1):131–6.
2. Ferner RE, Huson SM, Thomas N, Moss C, Willshaw H, Evans DG, *et al*. Guidelines for the diagnosis and management of individuals with neurofibromatosis 1. J Med Genet. 2007;44(2):81–8.
3. Fossali E, Minoja M, Intermite R, Spreafico C, Casalini E, Sereni F. Percutaneous transluminal renal angioplasty in neurofibromatosis. Pediatr Nephrol. 1995;9(5):623–5.
4. Friedman JM. Epidemiology of neurofibromatosis type 1. Am J Med Genet. 1999;89(1):1–6.
5. Friedman JM, Arbiser J, Epstein JA, Gutmann DH, Huot SJ, Lin AE, *et al*. Cardiovascular disease in neurofibromatosis 1: report of the NF1 Cardiovascular Task Force. Genet Med. 2002;4(3):105–11.
6. Oderich GS, Sullivan TM, Bower TC, Gloviczki P, Miller DV, Babovic-Vuksanovic D, *et al*. Vascular abnormalities in patients with neurofibromatosis syndrome type I: clinical spectrum, management, and results. J Vasc Surg. 2007;46(3):475–84.
7. Tedesco MA, Di Salvo G, Ratti G, Natale F, Calabrese E, Grassia C, *et al*. Arterial distensibility and ambulatory blood pressure monitoring in young patients with neurofibromatosis type 1. Am J Hypertens. 2001;14(6 Pt 1):559–66.
8. Corbetta JP, Duran V, Burek C, Weller S, Sager C, Dingevan R, *et al*. Renal autotransplantation for the treatment of renovascular hypertension in the pediatric population. J Pediatr Urol. 2011;7(3):378–82.
9. Eliason JL, Coleman DM, Criado E, Kershaw DB, Blatt NB, Williams DM, *et al*. Remedial operations for failed endovascular therapy of 32 renal artery stenoses in 24 children. Pediatr Nephrol. 2016;31(5):809–17.
10. Kim SS, Stein DR, Ferguson MA, Porras D, Chaudry G, Singh MN, *et al*. Surgical management of pediatric renovascular hypertension and midaortic syndrome at a single-center multidisciplinary program. J Vasc Surg. 2021;74(1):79–89.
11. Martinez A, Novick AC, Cunningham R, Goormastic M. Improved results of vascular reconstruction in pediatric and young adult patients with renovascular hypertension. J Urol. 1990;144(3):717–20.
12. Booth C, Preston R, Clark G, Reidy J. Management of renal vascular disease in neurofibromatosis type 1 and the role of percutaneous transluminal angioplasty. Nephrol Dial Transplant. 2002;17(7):1235–40.
13. Scott T, Venuthurupalli SK. Kidney autotransplantation as a treatment for resistant hypertension due to renal artery stenosis: a case report and review of the literature. Clin Nephrol Case Stud. 2022;10:1–5.
14. Stanley JC, Criado E, Upchurch GR Jr, Brophy PD, Cho KJ, Rectenwald JE, *et al*. Pediatric renovascular hypertension: 132 primary and 30 secondary operations in 97 children. J Vasc Surg. 2006;44(6):1219–28.
15. Coleman DM, Eliason JL, Beaulieu R, Jackson T, Karmakar M, Kershaw DB, *et al*. Surgical management of pediatric renin-mediated hypertension secondary to renal artery occlusive disease and abdominal aortic coarctation. J Vasc Surg. 2020;72(6):2035–46.
16. Jordan ML, Novick AC, Cunningham RL. The role of renal autotransplantation in pediatric and young adult patients with renal artery disease. J Vasc Surg. 1985;2(3):385–92.

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