

# Successful endovascular embolization of large symptomatic congenital renal arteriovenous fistula in an octogenarian

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## ABSTRACT

Congenital renal arteriovenous fistula (rAVF) is a rare and often underdiagnosed clinical condition. Here, we present a case of a large congenital rAVF in an 81-year-old woman with a right flank bruit and high-output heart failure. The rAVF was successfully treated with percutaneous endovascular coil embolization. Complications included a small right renal hematoma, mild contrast-induced nephropathy, and small right renal infarct in the lower pole. Postoperatively, the patient had complete resolution of symptoms with salvage of the kidney. She has been observed annually for 5 years with computed tomography scan and ultrasound examination. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:419-22.)

**Keywords:** Congenital renal arteriovenous fistula; Coil embolization; High-output heart failure; Renal hematoma

Renal arteriovenous fistulas (rAVFs) and renal arteriovenous malformations, rare pathologic connections between arteries and veins, occur in approximately 0.04% of the population.<sup>1-3</sup> They can be acquired, congenital, or idiopathic. Most (>70%) rAVFs are acquired as a result of iatrogenic injury, trauma, or neoplasm.<sup>1,2</sup> Congenital rAVFs are thought to be focal failures of embryologic vascular development.<sup>2</sup> Large, high-output rAVFs are often symptomatic, causing hypertension, hematuria, renal insufficiency, and heart failure exacerbations.<sup>4</sup> Whereas rAVFs are rare and underdiagnosed, advancements in imaging modalities have increased diagnosis and treatment.

Past treatments of large rAVFs have included nephrectomy and open ligation. However, an increasing number of cases have successfully undergone endovascular treatment with low morbidity and mortality.<sup>1,2</sup> This case describes successful endovascular embolization of a large congenital rAVF causing symptomatic congestive heart failure. The patient provided consent for publication of this report.

## CASE REPORT

An 81-year-old independent white woman with history of hypertension, hyperlipidemia, supraventricular tachyarrhythmia, mild aortic stenosis, mild mitral and tricuspid regurgitation, moderate pulmonary hypertension, insulin resistance, and

recurrent pleural effusions presented with progressive dyspnea on exertion during the past week. She additionally reported bilateral lower extremity edema, orthopnea, and occasional palpitations. On examination, she was tachypneic (respiratory rate of 30 breaths/min) with an oxygen saturation of 89% on room air, blood pressure of 165/76 mm Hg, and pulse of 95 beats/min. She appeared to be in moderate respiratory distress with decreased breath sounds, bilateral crackles, and use of accessory muscles. Cardiac examination revealed 2/6 systolic ejection murmur radiating to the carotids and 3/6 apical holosystolic murmur in the axilla. Abdominal examination revealed a loud right-sided abdominal bruit and thrill. She had palpable femoral pulses and pitting edema of both legs.

Laboratory testing revealed a white blood cell count of  $18.7 \times 10^9/L$ ; hemoglobin level, 13.0 g/dL; hematocrit, 40.0%; platelet count,  $254 \times 10^9/L$ ; and neutrophil count,  $12.0 \times 10^9/L$ . Pro-bone morphogenetic protein level was 2288 pg/mL; blood urea nitrogen concentration, 25 mg/dL; creatinine concentration, 1.2 mg/dL; and glomerular filtration rate, 28.7 mL/min/ $1.73 \text{ m}^2$ . Cardiac enzymes were negative. A chest radiograph demonstrated pulmonary congestion. Electrocardiography showed left anterior fascicular block but was otherwise normal. Echocardiography demonstrated reduced ejection fraction of 35%, severely elevated pulmonary artery systolic pressure, grade II diastolic dysfunction, and elevated left atrial pressure.

The patient was admitted and treated with ipratropium bromide-albuterol (DuoNeb) and bilevel positive airway pressure with symptomatic improvement. Her oxygen saturation improved to 94%, and the respiratory rate decreased to 16 breaths/min. She was treated with diuretics and continued to improve. An abdominal aortic ultrasound scan was obtained and demonstrated a large right rAVF with associated dilation of the right renal artery, vein, and inferior vena cava. Pulsatile flow was seen in the inferior vena cava. The cardiology service was consulted and believed that the rAVF was contributing significantly to her symptoms and warranted treatment. The vascular surgery service was consulted, and computed tomography angiography (CTA) was obtained; it demonstrated a dilated right renal artery (11 mm), dilated renal vein (27 mm), and large right rAVF (Figs 1 and 2). There were four renal artery branches

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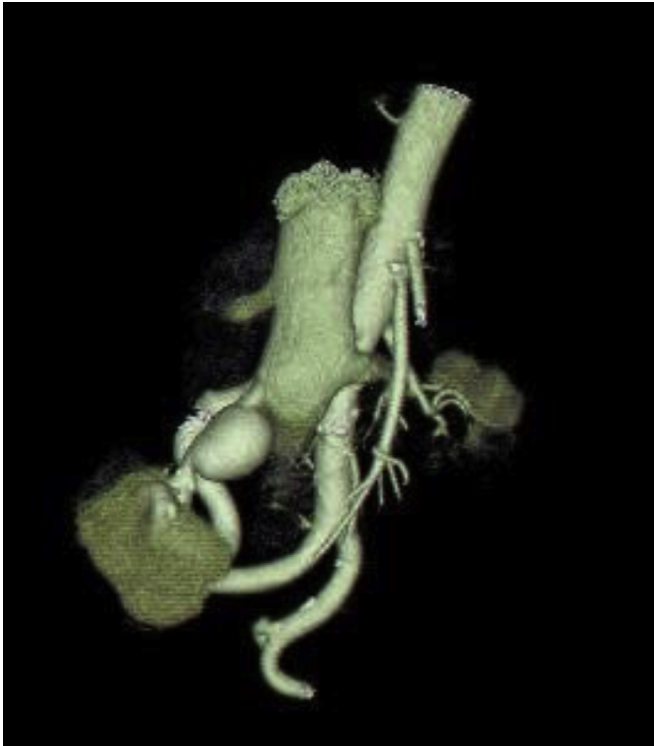
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**Fig 1.** Preoperative computed tomography angiography (CTA) image reconstruction demonstrating renal arteriovenous fistula (rAVF).



**Fig 2.** Preoperative computed tomography angiography (CTA) image demonstrating dilated right renal artery (*thin arrow*) and dilated and contrast-enhanced right renal vein (*thick arrow*).

proximal to the rAVF. The patient denied any history of trauma, surgery, or instrumentation. Endovascular repair was recommended. Stent graft placement was considered but decided against, given the tortuosity of the renal artery; thus, percutaneous embolization of the right rAVF was recommended with the understanding that a portion of the kidney would become ischemic.



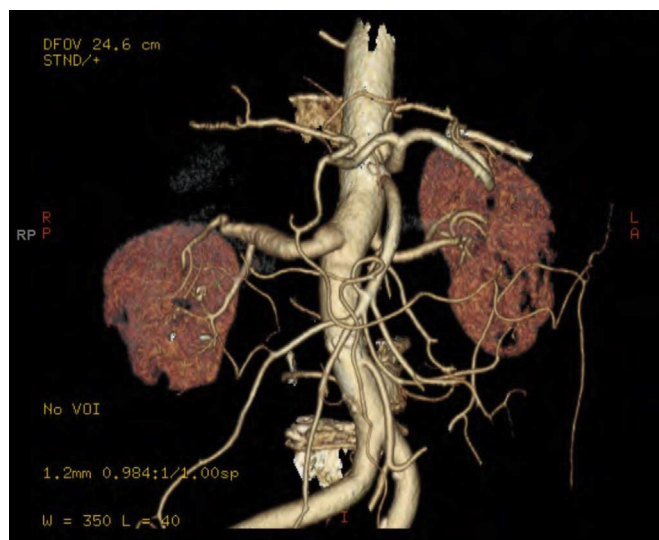
**Fig 3.** Diagnostic right renal artery angiogram with renal arteriovenous fistula (rAVF).

Following medical optimization, the patient underwent percutaneous embolization of the right rAVF. Right femoral access was obtained and secured with a 5F sheath. A 0.035-inch Glidewire (Terumo Interventional Systems, Somerset, NJ) and rim catheter were used to cannulate the right renal artery. The Glidewire was exchanged for a stiff Glidewire, and an 8F sheath was advanced to the distal renal artery. Angiography demonstrated four patent renal branches and the rAVF beyond these branches (Fig 3). A 14-mm Amplatzer plug (Abbott, St. Paul, Minn) was deployed in the distal renal artery, distal to the branches. Angiography showed significantly decreased flow in the rAVF (Fig 4). The access site was closed with Perclose ProGlide (Abbott Vascular, Santa Clara, Calif). A total of 120 mL of contrast material was used. She became hypotensive postoperatively. CTA demonstrated a small hematoma around her right kidney with subsequent drop in hemoglobin level from 13.1 g/dL to 11.1 g/dL. She responded well to a blood transfusion and gentle hydration. She complained of mild right flank pain postoperatively that was controlled with pain medication. Vital signs and hemoglobin level were stable, and her pain resolved. Creatinine concentration increased to 1.5 mg/dL but decreased to 1.3 mg/dL (baseline, 1.2 mg/dL). Glomerular filtration rate was 26 mL/min/1.73 m<sup>2</sup> on admission and 24 mL/min/1.73 m<sup>2</sup> at discharge. Electrolytes were stable, and she was discharged on postoperative day 2.

CTA performed 1 week postoperatively demonstrated successful occlusion of the rAVF as well as infarction of the lower pole of the right kidney (Fig 5). The remaining portion of the kidney showed normal enhancement. After discharge, the patient saw her primary care provider, who doubled the dose of furosemide. Laboratory evaluation performed after CTA demonstrated



**Fig 4.** Completion angiogram after embolization.



**Fig 5.** Follow-up computed tomography angiography (CTA) image showing resolution of the renal arteriovenous fistula (rAVF).

significant increases in creatinine concentration to 4.1 mg/dL, in potassium concentration to 7.3 mmol/L, in sodium concentration to 129 mmol/L, and in hemoglobin level to 10.5 mg/dL. She was admitted for contrast-induced nephropathy exacerbated by excessive diuresis, hyperkalemia, and hyponatremia. The nephrology service was consulted. She responded well to medical management and did not require dialysis. Her creatinine concentration decreased to 1.7 mg/dL, and she was discharged on hospital day 3. The patient continued to do well

with complete resolution of her congestive heart failure symptoms, and her renal function returned to baseline. She has been observed annually with renal artery duplex ultrasound examination for the past 6 years.

## DISCUSSION

The rAVF is a rare entity and is usually congenital or acquired (iatrogenic, traumatic, or malignant). Congenital rAVFs are classified by type or appearance. The cirrloid type appears with a webbing of multiple varices, whereas idiopathic rAVFs appear to have one or two large cavernous connections with well-defined arterial and venous components.<sup>4,5</sup> Idiopathic rAVFs represent only 2% to 5% of all rAVFs.<sup>6</sup> In the absence of trauma or instrumentation, our patient was diagnosed with a congenital idiopathic rAVF. Typically, small rAVFs are asymptomatic; however, larger ones result in hypertension, hematuria (50% of cases), flank pain, congestive heart failure, and occasionally a palpable abdominal mass.<sup>4</sup> Our patient presented with congestive heart failure and an abdominal bruit, prompting imaging that revealed the rAVF.

Improvements in noninvasive imaging have allowed increased diagnosis and treatment of rAVFs.<sup>1</sup> Duplex ultrasound is an effective screening modality for patients with a suspected rAVF.<sup>6</sup> Once the diagnosis is confirmed, CTA is typically performed to guide treatment. Catheter-directed renal angiography is considered the “gold standard” for diagnosis of rAVFs.<sup>2</sup> In our patient, ultrasound confirmed the rAVF, but CTA was necessary for surgical planning.

Treatment of rAVF ranges from surveillance for asymptomatic patients to surgical intervention for symptomatic patients, with endovascular and open options. The decision to treat and the treatment approach should be based on symptoms, age of the patient, blood pressure, comorbidities, and renal function. It is important to consider the complexity of repair required, with attention to vascular anatomy, available technology, and operator experience.<sup>1</sup> Our patient was an independent 81-year-old woman with high-output heart failure due to the rAVF. Treatment was recommended. Surgical options include open removal of an associated aneurysm (if present) as well as ligation of inflow and outflow vessels, ex vivo repair with autotransplantation, and even nephrectomy.<sup>1,7</sup> Alternative treatment with endovascular techniques, including coil embolization and stent graft placement, have been described. The preferred endovascular approach for treatment of congenital rAVF is coiling.<sup>1,3</sup>

Complications of coiling include arterial dissection, worsening renal function, coil migration, and pulmonary embolus.<sup>1,3</sup> Decreased renal function results from exposure to contrast material or partial loss of parenchyma

after coil embolization.<sup>1,2</sup> Unfortunately, an estimated <25% loss of parenchyma during these procedures is inevitable; however, this does not typically affect overall renal function.<sup>2</sup> Embolization also confers a risk of coil migration into an already dilated inferior vena cava and even into the pulmonary circulation.<sup>1-3</sup> Prevention methods include use of a partially deployed wall stent, vascular plugs, and the “stop-flow” technique with occlusion balloons.<sup>2,3</sup>

## CONCLUSIONS

Congenital rAVFs represent only a small component of all vascular abnormalities. They are often asymptomatic but can cause significant symptoms, including hypertension, flank pain, congestive heart failure, and hematuria. Symptomatic rAVFs typically require surgical intervention through open or endovascular methods. Endovascular techniques using embolization have become a successful treatment option with decreased surgical risks. Postoperative surveillance is recommended, however, as long-term data regarding success are not available.

## REFERENCES

1. Zhang Z, Yang M, Song L, Tong X, Zou Y. Endovascular treatment of renal artery aneurysms and renal arteriovenous fistulas. *J Vasc Surg* 2013;57:765-70.
2. Nassiri N, Dudy Y, Carroccio A, Rosen R. Transarterial treatment of congenital renal arteriovenous fistulas. *J Vasc Surg* 2013;58:1310-5.
3. Thatigotla BC, Mahalingam S, Cosgrove JM. Congenital giant renal arteriovenous fistula presenting as congestive heart failure. *Vasc Dis Manag* 2008;5:180-2.
4. Perkov D, Novacic K, Novosel L, Knezevic N. Percutaneous embolization of idiopathic renal arteriovenous fistula using Amplatzer vascular plug II. *Int Urol Nephrol* 2013;45:61-8.
5. Kopchick JH, Bourne NK, Fine SW, Jacobsohn HA, Jacobs SC, Lawson RK. Congenital renal arteriovenous malformations. *Urology* 1981;17:13-7.
6. Paschalis-Purtak K, Januszewicz M, Rokicki A, Pucilowska B, Imiela J, Cybulska I, et al. Arteriovenous fistula of the kidney: a case report of 47-year-old female patient treated by embolization. *J Hum Hypertens* 2003;17:293-6.
7. Cho KJ, Stanley JC. Non-neoplastic congenital and acquired renal arteriovenous malformations and fistulas. *Radiology* 1978;129:333-43.

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