

Successful Renal Artery Stenting in an Infant With Middle Aortic Syndrome: Resolution of Malignant Hypertension and Normalization of Renal Growth

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Abstract

Renal artery stenosis (RAS) in infants is rare but often results in severe hypertension and renal failure. Middle aortic syndrome (MAS) is a common etiology of RAS in children. Management remains controversial due to limited evidence defining optimal therapy in infants. Vascular stenting in infants is limited by anatomical and technical challenges. We present a case of renal artery stenting in an infant with MAS. The patient presented neonatally with malignant hypertension and renal hypoperfusion. Progressive right RAS and persistent hypertension required multiple antihypertensive agents. Surgery was initially not feasible because of the patient's size. At 22 months, the patient underwent successful renal artery stenting followed by balloon dilation. Following intervention, blood pressure normalized, and right kidney growth recovered. This case illustrates that renal artery stenting can result in sustained resolution of hypertension and normalization of renal growth in infants with severe MAS when surgery is not feasible.

Summary

What is new

First reported renal artery stenting in an infant with middle aortic syndrome achieving medication-free normotension and normalization of renal growth.

Introduction

Renal artery stenosis (RAS) is a rare but important cause of severe hypertension and kidney injury in children [1]. Middle aortic syndrome (MAS) and Williams syndrome are among the most common causes of pediatric RAS [2, 3]. Management remains challenging because evidence guiding treatment in infants is limited [4]. Treatment options include medical therapy, surgical revascularization, and endovascular intervention [2]. Vascular stenting is rarely performed in infants because of small vessel size, concerns regarding somatic growth, and limited long-term outcome data [5]. However, emerging evidence suggests favorable outcomes with stent angioplasty in selected pediatric patients in comparison to angioplasty alone [6].

We report an infant with MAS-associated critical right RAS who underwent successful renal artery stenting at 22 months, resulting in resolution of hypertension, discontinuation of antihypertensive medications, and recovery of renal growth. This case illustrates that renal artery stenting can achieve excellent clinical outcomes in infants with severe MAS when surgical treatment is not feasible.

Case Presentation

DL presented during the first week of life with recalcitrant hypertension (92–120 mmHg systolic and 56–89 mmHg diastolic) and evidence of right renal hypoperfusion on ultrasound with increased cortical echogenicity. Echocardiography demonstrated left ventricular dysfunction of 45% without coarctation of

the aorta. Serum creatinine was elevated (peak 0.88 mg/dL). Magnetic resonance imaging at 10 days of age demonstrated significant hypoplasia of the abdominal aorta extending from the diaphragm to the iliac bifurcation, measuring 3-4mm in diameter. Magnetic resonance angiography at 2 weeks of age identified diffuse narrowing of the abdominal aorta measuring approximately 3 mm and diffusely attenuated renal arteries, confirming the diagnosis of middle aortic syndrome (MAS).

The patient was initially managed with intravenous nitroprusside, esmolol, and hydralazine and later transitioned to oral clonidine, captopril, and amlodipine. Regional and national experts favored medical management because surgical repair was considered high risk with low potential success given the patient's age and size. Renal function normalized by 3 months, and he remained clinically well without intestinal or lower extremity ischemia. However, serial imaging demonstrated progressive right RAS with impaired right kidney growth, while hypertension persisted despite treatment with three antihypertensive agents.

At 14 months of age, computed tomographic angiography showed severe focal narrowing of the right renal artery, mild left RAS, and persistent diffuse hypoplasia of the abdominal aorta. Following multidisciplinary discussion, the family elected to pursue endovascular intervention. At 22 months of age, catheter angiography confirmed critical right RAS with delayed distal filling (Fig. 1A). The lesion was crossed with a coronary guidewire and predilated using a 1.5 x 8 mm coronary balloon (Fig. 1B). Placement of a 2.25 x 12 mm everolimus-eluting stent was then deployed in the proximal right renal artery and expanded to 2.6 mm (Fig. 1C-D). Completion angiography demonstrated a patent renal artery without residual stenosis (Fig. 1E). The procedure was well tolerated without complications.

Following intervention, blood pressure normalized and antihypertensive medication were gradually discontinued. Four months later, surveillance imaging suggested in-stent restenosis. Repeat angiography at 27 months confirmed moderate in-stent stenosis, which was successfully treated with high-pressure balloon angioplasty (Fig. 1F-H).

After repeat intervention, renal artery flow normalized and antihypertensive therapy was completely discontinued. Serial renal ultrasounds demonstrated recovery of right kidney growth to normal size, no residual right RAS, and only middle residual left RAS. At the last visit, when the patient was 5 years old, blood pressure remained normal without medication, renal growth was normal bilaterally, and the patient remained asymptomatic.

Discussion

We report the successful management of an infant with MAS and critical RAS who presented neonatally with malignant hypertension, renal hypoperfusion, and left ventricular dysfunction. Despite limited treatment options because of his age and size, he achieved complete resolution of hypertension, recovery of renal growth, and discontinuation of all antihypertensive medications following endovascular intervention.

Management of pediatric renovascular disease is particularly challenging in infants [8].

Medical therapy is typically first-line because surgical revascularization is technically difficult in young children, given small vessel caliber and the diffuse nature of vascular involvement [7]. In our patient, multiple national referral centers concluded that surgery carried substantial risk with uncertain benefit, leaving endovascular therapy as the primary opportunity to restore renal perfusion and prevent renal atrophy. Stenting is generally avoided due to small vessel caliber, somatic growth, in-stent restenosis, and limited long-term outcome data [2, 5].

To our knowledge, this patient is among the youngest reported to undergo renal artery stenting for MAS-associated RAS [6, 8]. Coronary drug-eluting stents have been an established option for small-caliber arteries in adults, and used in pediatric renal vessels below 4 mm diameter [9]. Successful deployment in this infant demonstrates the feasibility of coronary interventional platforms to extend endovascular therapy to infant-sized renal vessels.

Beyond blood pressure control, recovery of renal growth is an important finding. Before intervention, serial imaging demonstrated progressive renal asymmetry and impaired growth despite medical therapy. Following restoration of renal perfusion, kidney growth normalized and renal size became comparable to the contralateral kidney. Although long-term renal function remains to be determined, this observation suggests that timely revascularization may permit recovery of renal development even after growth impairment has occurred.

Previous reports have favored angioplasty alone because of concerns regarding restenosis and future vessel growth [2]. In the largest cohort of children younger than two years, RAS was managed predominantly by angioplasty, with stenting essentially avoided and surgery reserved for angioplasty or medical failure [8]. Our patient developed moderate in-stent restenosis within four months, but repeat balloon angioplasty successfully restored vessel patency and maintained favorable outcomes. This supports a staged approach in which repeat angioplasty may address restenosis and accommodate future somatic growth.

In summary, renal artery stenting can provide excellent clinical outcomes in selected infants with severe MAS when surgery is not feasible. Medical therapy served as a bridge to delayed definitive revascularization [10]. Medication-free normotension is notable because most children with RAS and MAS remain hypertensive after intervention [4]. Although long-term surveillance remains necessary, successful stent implantation, re-dilation, medication-free normotension, and recovery of renal growth suggest that endovascular intervention may be a valuable option in carefully selected patients.

Declarations

Conflict of Interest Disclosures (includes financial disclosures): The authors have no conflicts of interest to disclose.

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Consent for publication: The patient's parent has consented to the publication of this case report and any accompanying images.

Clinical Trial Registration (if any): Not applicable.

Abbreviations: RAS – Renal Artery Stenosis; MAS – Middle Aortic Syndrome; MRI – Magnetic Resonance Imaging; CTA – Computed Tomography Angiography.

Article Summary

A novel case demonstrating successful renal artery stenting in an infant with middle aortic syndrome, resolving malignant hypertension and promoting renal growth.

Contributors Statement Page

Ms. Nicole Wong reviewed the case and the literature, and drafted the manuscript.

Dr. Andras Bratincsak supervised patient care, conceptualized the case report, and contributed to the manuscript preparation.

Both authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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Figures

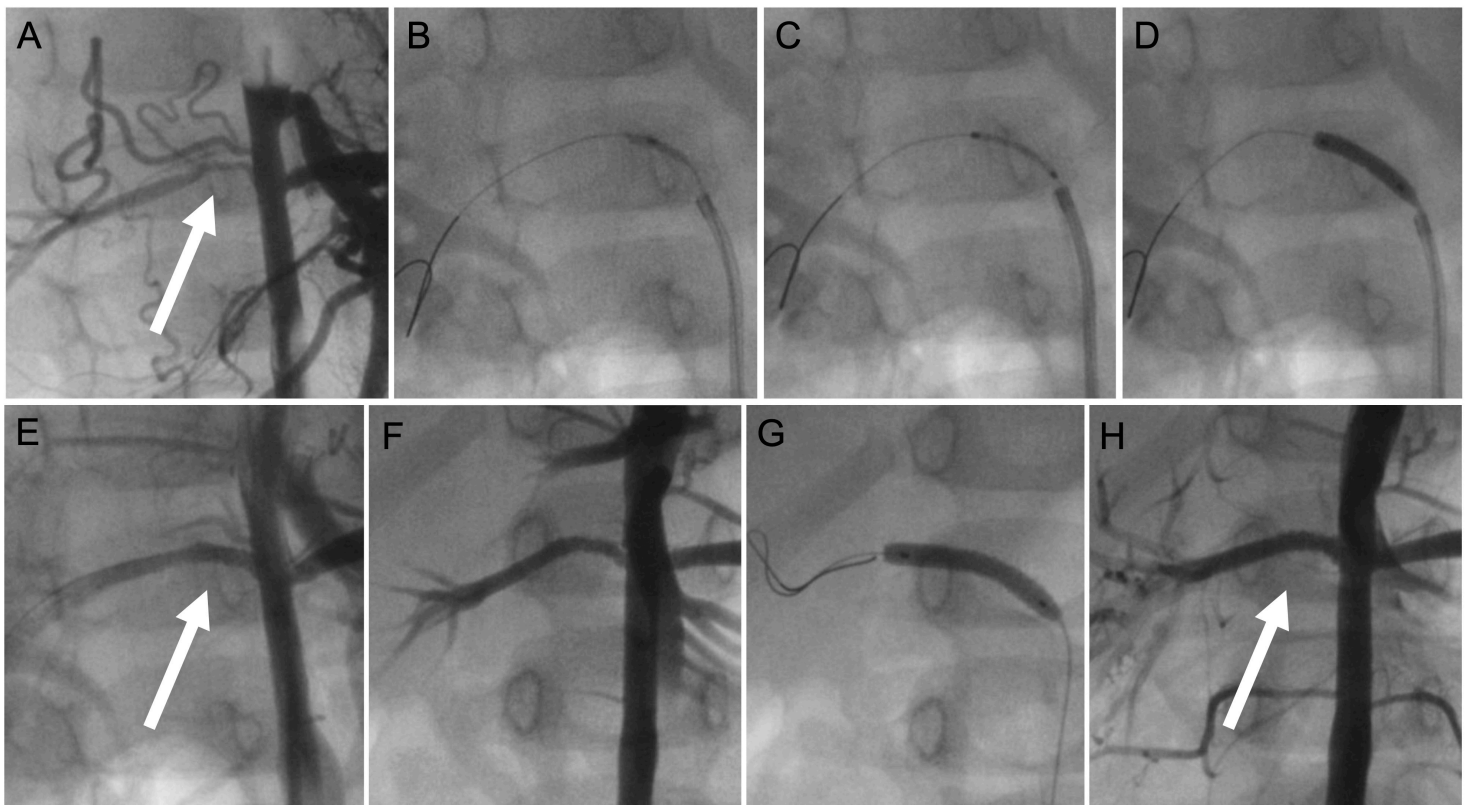


Figure 1

Balloon angioplasty and stent placement in the right renal artery with excellent results. 1A: Initial angiogram demonstrating severe proximal stenosis of the right renal artery (white arrow). 1B: Balloon angioplasty with a 1.5 mm coronary balloon. 1C: Coronary artery stent inserted in the dilated proximal segment. 1D: Balloon dilation with the coronary artery stent to 2.25 mm. 1E: Angiogram after stent placement demonstrated an improved proximal renal artery and better distal filling. 1F: Repeat catheter angiogram 4 months after the stent placement showed mild to moderate in-stent re-stenosis and narrowed diameter of the proximal right renal artery. 1G: Balloon angioplasty with a coronary artery balloon dilated the stent up to 3.6 mm. 1H: Angiogram after balloon dilation demonstrated a much improved, now normal diameter of the right renal artery without any residual stenosis