

Idiopathic midaortic syndrome

David Saul¹, Rahul Nikam¹, Vinay Kandula¹, Achala Donuru²

¹Department of Medical Imaging, Nemours Alfred I. DuPont Hospital for Children, Wilmington, DE 19803, USA, ²Department of Radiology, Thomas Jefferson University Hospitals, Philadelphia, PA 19107, USA

ABSTRACT

Midaortic syndrome (MAS) is characterized by narrowing of the descending aorta between the distal aortic arch and the aortic bifurcation. We present the case of a 4-year-old male presenting with a murmur and diagnosed with MAS. He was treated with a thoracoabdominal bypass graft.

Keywords: Aortic bypass surgery, aortic coarctation, renovascular hypertension

CLINICAL SUMMARY

A 4-year-old boy was noted to have a murmur by his pediatrician. He was referred to a cardiologist, who measured his blood pressure at 160/60. Echocardiogram revealed concentric left ventricular hypertrophy (LVH) with a “drag” in the descending thoracic aorta consistent with coarctation. Computed tomography (CT) angiogram of the abdomen revealed an extremely narrow upper abdominal aorta [Figure 1a and b], from which a diagnosis of midaortic syndrome (MAS) was made. There was no soft-tissue thickening surrounding the narrowed portion (as is often seen in Takayasu arteritis), nor was there involvement of branch vessels to specifically suggest other vasculopathy such as fibromuscular dysplasia as a cause. There was no clinical evidence of neurofibromatosis or Williams syndrome.

The patient was started on four blood pressure medications (beta-blocker - atenolol, vasodilator - minoxidil, angiotensin-converting enzyme inhibitor - enalapril, and calcium channel blocker - amlodipine), with blood pressure reduction to 120s/50s in the arms and 40s/20s in the legs. The patient was eventually unable to walk due to severe claudication and was referred to a tertiary hospital for a

thoracoabdominal bypass graft. Polytetrafluoroethylene graft was successfully placed from the level of the diaphragm to just above the origin of the inferior mesenteric artery [Figure 2]. Postoperatively, his blood pressure was controlled with one antihypertensive medication (amlodipine). On follow-up echo 18 months later, LVH had resolved. He is now completely asymptomatic with no complaints of chest pain, dizziness, shortness of breath, syncope, or fatigue.

Differential diagnosis

Takayasu arteritis, neurofibromatosis type 1, fibromuscular dysplasia.

DISCUSSION

MAS is characterized by narrowing of the descending aorta between the distal aortic arch and the aortic bifurcation, comprising 0.5%–2% of cases of aortic stenosis.^[1] Disease involvement may be focal or long segment smooth narrowing and can extend into visceral arterial branches. Focal stenosis most commonly occurs in the suprarenal portion (60%–70% of cases), with focal renal (20%–25%) and infrarenal (10%–15%) involvement being less frequent.^[2]

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Saul D, Nikam R, Kandula V, Donuru A. Idiopathic midaortic syndrome. *Ann Pediatr Card* 2021;14:561-3.

Access this article online	
Quick Response Code: 	Website: www.annalspc.com
	DOI: 10.4103/apc.apc_261_20

Address for correspondence: Dr. Achala Donuru, Department of Radiology, Thomas Jefferson University Hospitals, 132 S 10th Street, 1079 Main Building, Philadelphia, PA 19107, USA.

E-mail: achala.donuru@jefferson.edu

Submitted: 19-Nov-2020

Revised: 24-Feb-2021

Accepted: 01-Mar-2021

Published: 25-Mar-2022

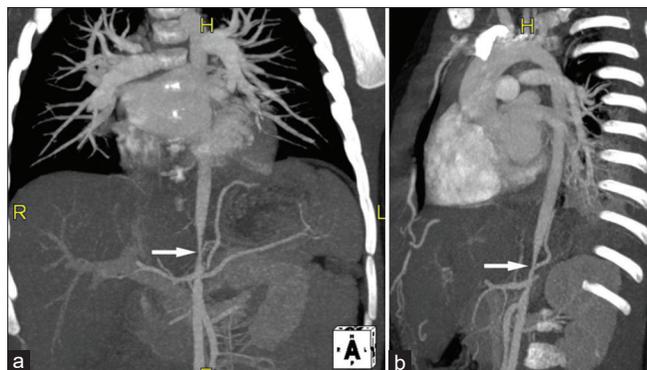


Figure 1: (a and b) Coronal and sagittal maximum intensity projection images demonstrate narrowing of the upper abdominal aorta (arrow)

Patients with MAS present predominantly with refractory hypertension. If left untreated, symptoms can progress to intermittent claudication of the legs, congestive heart failure, and renal insufficiency. Symptoms include sequelae of hypertension, renal insufficiency, mesenteric ischemia, solid organ hypoperfusion, and lower limb ischemia. Chronic renal ischemia leading to renovascular disease can further exacerbate refractory hypertension. Untreated MAS has a 90% mortality rate by the sixth decade of life due to end-organ damage.

MAS can be associated with inflammatory arteritis including Takayasu's, neurofibromatosis type 1, fibromuscular dysplasia, mucopolysaccharidosis, congenital Rubella syndrome, Noonan's syndrome, and Williams syndrome.

MAS may be difficult to diagnose clinically, as it is one of the rarest causes of refractory hypertension. Renal function laboratory values may be abnormal, which could prompt further evaluation. Echocardiography and renal ultrasound can be useful initial imaging tests to evaluate for thoracic coarctation and renal parenchymal atrophy. While not definitive, both of these examinations may show secondary or direct evidence of aortic narrowing. Angiography was formerly the gold standard to diagnose MAS, but definitive diagnosis is now often achieved with either CT or magnetic resonance angiography.

The treatment of mid-aortic syndrome can be medical, endovascular, or surgical. A combination of medical management and invasive strategies can result in adequate control of hypertension and preservation of end-organ function.

Percutaneous interventions are effective in relieving obstruction in the acute setting, but there is a high incidence of refractory hypertension and recurrent stenosis requiring repeat intervention. Up to 50% of endovascular repairs require reintervention within 5 years of surgery.^[3] Serial dilation utilizing drug-eluting balloons may have promise as a percutaneous option.



Figure 2: Volume-rendered image from a computed tomography angiogram of the chest and abdomen in the postoperative period demonstrates narrowing of the upper abdominal aorta (arrow) and the polytetrafluoroethylene graft (arrow head)

Percutaneous interventions can be associated with severe complications, including vascular tears, development of aneurysms, dissection, and even death.^[4]

Surgical interventions include aorto-aortic bypass, patch aortoplasty, and primary aortic repair. Improvement or resolution of hypertension after surgery in the mid to long term has been reported at 70%.^[5] Freedom from reintervention after surgery is reported to be 72% at 10 years.^[2] Complications occur in 9% of cases and include graft stenosis, bleeding, thrombosis, and iatrogenic tears. The mortality rate is 2.9%–4%.^[6]

While rare, MAS is a highly morbid and poorly understood disease process that occurs in both children and adults. The timing of disease progression is variable, but the risk of end-organ damage and death remains high no matter the age of presentation. It is important for imagers and clinicians to be aware of this disease and to have a high index of suspicion on screening. This can facilitate early diagnosis and treatment, as management is often a stepwise process that may require both noninvasive and surgical approaches.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Yıldızdaş HY, Erdem S, Demir F, Şimşek H, Özlü F, Ozbarlas N. An infant with congenital midaortic syndrome. *Cukurova Med J* 2018;43:1042-44.
2. Porras D, Stein DR, Ferguson MA, Chaudry G, Alomari A, Vakili K, et al. Midaortic syndrome: 30 years of experience with medical, endovascular and surgical

- management. *Pediatr Nephrol* 2013;28:2023-33.
3. Patel RS, Nguyen S, Lee MT, Price MD, Krause H, Truong VT, *et al.* Clinical Characteristics and Long-Term Outcomes of Midaortic Syndrome. *Ann Vasc Surg* 2020;66:318-25.
 4. Rao SA, Mandalam KR, Rao VR, Gupta AK, Joseph S, Unni MN, *et al.* Takayasu arteritis: Initial and long-term follow-up in 16 patients after percutaneous transluminal angioplasty of the descending thoracic and abdominal aorta. *Radiology* 1993;189:173-9.
 5. Taketani T, Miyata T, Morota T, Takamoto S. Surgical treatment of atypical aortic coarctation complicating Takayasu's arteritis--experience with 33 cases over 44 years. *J Vasc Surg* 2005;41:597-601.
 6. Rumman RK, Nickel C, Matsuda-Abedini M, Lorenzo AJ, Langlois V, Radhakrishnan S. Disease beyond the arch: A systematic review of middle aortic syndrome in childhood. *Am J Hypertens* 2015;28:833-46.