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Mid-aortic syndrome diagnosed by transesophageal echocardiogram in a patient with dilated cardiomyopathy

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Abstract

Mid-aortic syndrome is a rare condition characterised by segmental narrowing of the thoracoabdominal aorta. Here, we demonstrate a case of mid-aortic syndrome in a 30-month-old female who was diagnosed via transesophageal echocardiography after presenting with dilated cardiomyopathy and severe heart failure requiring placement of a left ventricular assist device.

Case presentation

Mid-aortic syndrome is a rare disorder characterised by narrowing of the thoracoabdominal aorta that most commonly presents with hypertension. This is typically diagnosed by angiography.[1]

Here, a 30-month-old female presented with cardiomegaly and was diagnosed with dilated cardiomyopathy without identified infectious or genetic cause. Heart failure medications were initiated but, due to clinical deterioration, she was eventually listed for cardiac transplantation and underwent placement of a left ventricular assist device as a bridge. Post-operatively, her central venous pressure was elevated, raising concerns for right ventricular dysfunction. Transthoracic windows were limited, so a transesophageal echocardiogram was performed which demonstrated normal right ventricular function. Due to a newly noted upper to lower extremity blood pressure discrepancy, a non-standard transesophageal echocardiography view was performed by advancing the probe into the stomach and orienting it posteriorly at a 90-degree angle which demonstrated severe narrowing of the abdominal aorta with an obstructive Doppler pattern (Fig 1). Catheterisation revealed severe narrowing of the abdominal aorta and renal arteries (Fig 2) with a 50-mmHg peak-to-peak pressure gradient. The patient underwent polytetrafluoroethylene graft placement around the stenotic region with bilateral renal artery reconnection distal to the stenosis (Fig 3). She was subsequently decannulated from the device and later removed from the transplant list. Six years later, she continues to do well on oral heart failure medications.

In this first reported diagnosis of mid-aortic syndrome by transesophageal echocardiography, evaluation by non-standard imaging allowed for surgical repair and clinical recovery. This case also highlights that this rare diagnosis may be initially missed by transthoracic echocardiography as the abdominal aorta is not typically evaluated and should be considered with the presentation of both cardiomyopathy and hypertension.

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Conflicts of interest. None.

Reference

1. Rumman RK, Nickel C, Matsuda-Abedini M, et al. Disease beyond the arch: a systematic review of middle aortic syndrome in childhood. Am J Hypertens 2015; 28:833–846.

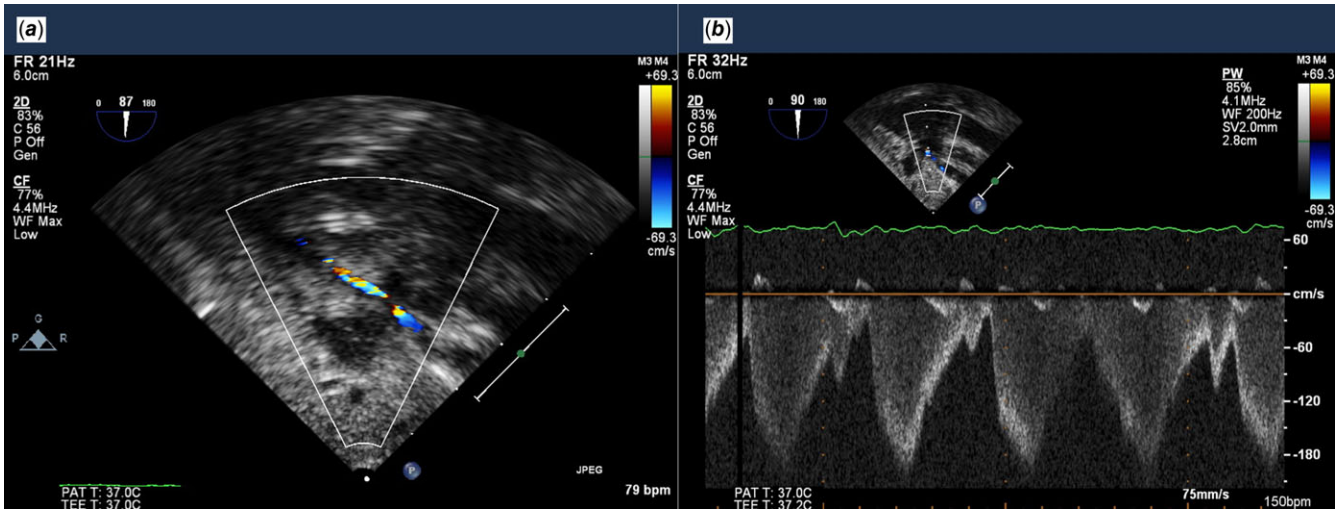

Figure 1. (***a***) Non-standard transesophageal echocardiography with the probe advanced into the stomach and oriented posteriorly at a 90-degree angle demonstrating severe narrowing of the abdominal aorta with aliasing of flow. (***b***) Pulsed wave Doppler of flow through the narrowed abdominal aorta demonstrating continuous antegrade flow in an obstructive pattern.

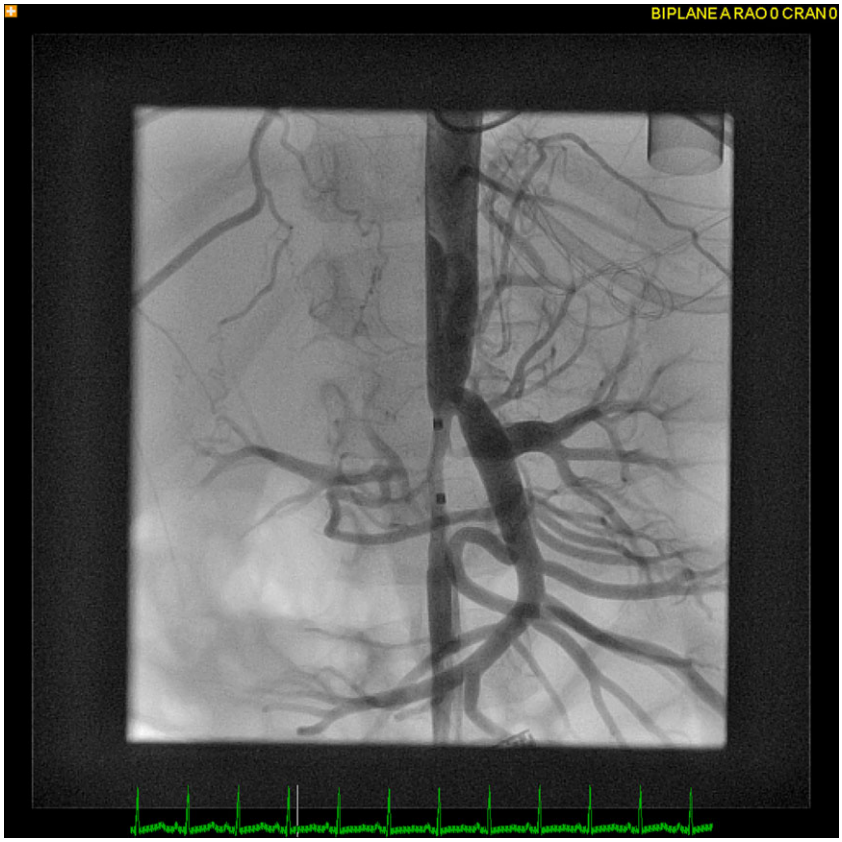

Figure 2. Angiography of the abdominal aorta demonstrating severe narrowing of the abdominal aorta just below the origin of the superior mesenteric artery with associated severe narrowing of the origins and proximal courses of the renal arteries.

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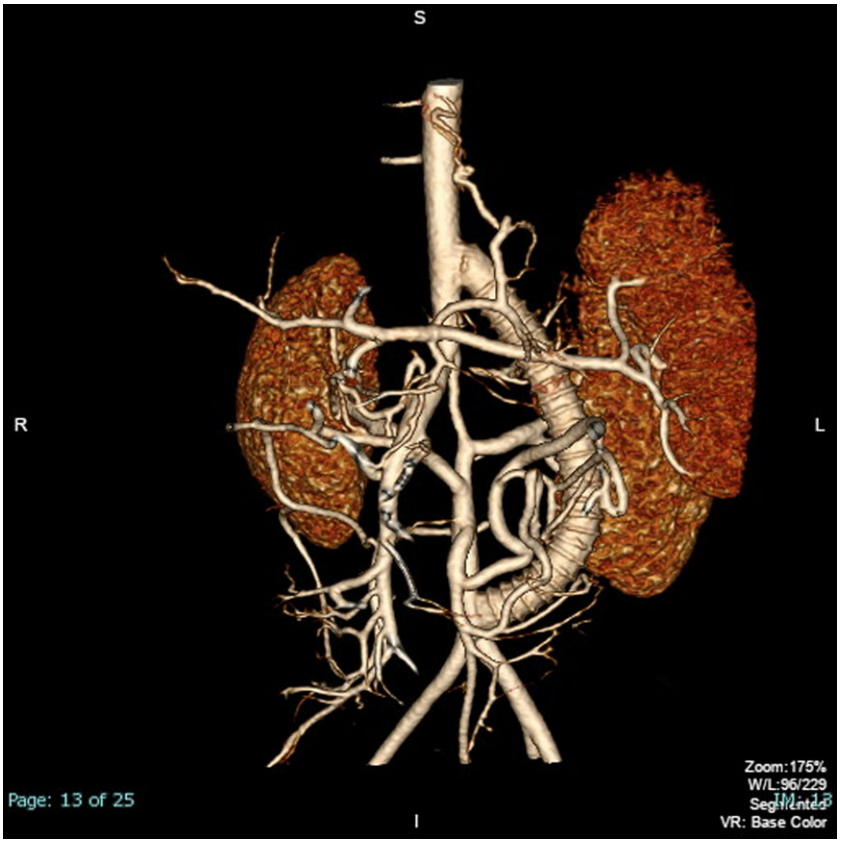

Figure 3. 3-dimensional reconstruction of computed tomography angiography after repair demonstrating a polytetrafluoroethylene graft around the stenotic region and normal caliber renal arteries.

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