Cardiology in the Young

cambridge.org/cty

Infantile dilated cardiomyopathy due to congenital left main coronary ostial stenosis

Jacqueline Doyle, Tammy Churchill and Shahryar M. Chowdhury

Department of Paediatrics, Division of Paediatric Cardiology, Medical University of South Carolina, Charleston, SC, USA

Brief Report

Cite this article: Doyle J, Churchill T, and Chowdhury SM (2021) Infantile dilated cardiomyopathy due to congenital left main coronary ostial stenosis. *Cardiology in the Young* **31**: 130–131. doi: 10.1017/S1047951120003224

Received: 29 April 2020
Revised: 27 August 2020
Accepted: 11 September 2020
First published online: 13 October 2020

Keywords:
Congenital left main coronary ostial stenosis; coronary anomaly; dilated cardiomyopathy

Author for correspondence:
Shahryar M. Chowdhury, MD, MSCR, 10 McClellan Banks Dr, MSC 915, Charleston, SC 29425, USA. Tel: +1 843 876 2273; Fax: +1 843 792 5878.
E-mail: chowdhur@musc.edu

Abstract

Although a rare form of congenital heart disease, anomalies of the coronary arteries can present as heart failure in infants. The most common lesion is an anomalous left coronary artery arising from the pulmonary artery, but other abnormalities can present similarly. This case is an infant who is found to have left coronary ostial stenosis causing dilated cardiomyopathy.

Congenital anomalies of the coronary arteries are a rare form of congenital heart disease. They have been reported to present at any age of life. In an infant, coronary artery anomalies can present with cardiac ischaemia leading to dilated cardiomyopathy and resultant failure to thrive. We report on a rare case of congenital left main coronary ostial stenosis that is presented as failure to thrive.

Case

A 4-month-old male who was born at 34 weeks was transferred to our hospital after a 3-week stay at an outside hospital for failure to thrive. He was transferred due to parental request for a second opinion on having a gastrostomy tube placed. Echocardiography during his initial workup at the outside hospital showed mild dilated cardiomyopathy with a left ventricular ejection fraction of 49%. Medical treatment with enalapril and furosemide was initiated.

Upon transfer, repeat echocardiogram at our hospital revealed severe dilated cardiomyopathy with a left ventricular ejection fraction of 24%, moderate mitral regurgitation and a small pericardial effusion. The coronary arteries were noted to have originated from their appropriate sinuses. Family history was negative for cardiomyopathy. Milrinone was started and the evaluation process for candidacy for heart transplantation was started.

A week later, ST-segment elevation was noted on telemetry. Review of the initial echocardiogram was concerning for left main coronary ostial stenosis due to aliasing of the color Doppler signal at the left main coronary artery origin (Fig 1a and Supplementary video S1). The next day, computed tomographic angiography also showed left main coronary artery stenosis (Fig 1b). A troponin I was 1.33 ng/ml afterward and ST-segment elevation was noted on the electrocardiogram. For further characterisation of the anatomy and haemodynamic assessment, the patient was taken to the cardiac catheterisation laboratory where his pulmonary capillary wedge pressure was 15 mmHg and angiograms confirmed the diagnosis of severe left main coronary artery stenosis (Fig 2 and Supplementary video S2).

The patient was taken to the operating room, severe congenital left main ostial stenosis was found. Repair was performed with a pericardial patch over the left main coronary ostium. His mitral valve was left unrepaired. The post-operative course was uncomplicated. An echocardiogram on post-op day 12, the day of discharge, showed objective improvement of left ventricular function with an ejection fraction of 44%. Left ventricular function continued to improve. At 4-year follow-up, the patient was growing well, was asymptomatic, and the left ventricular ejection fraction was 58%. The left coronary artery was widely patent (Fig 1c). He did exhibit moderate–severe mitral regurgitation and worsening left ventricular dilation, consideration is ongoing regarding the timing of returning to the operating room for mitral valve repair.

Discussion

Dilated cardiomyopathy caused by a congenital anomaly of the coronary vasculature in infancy can be caused by a normally positioned coronary origin that exhibits ostial atresia or stenosis, or, more commonly, can be caused by an anomalous coronary artery arising from the pulmonary artery.[1] This was a rare case of congenital left main coronary artery stenosis that led to significantly reduced left ventricular function and failure to thrive early in life.

Reports of left main coronary artery stenosis are often associated with other congenital anomalies, such as coarctation of the aorta, truncus arteriosus and supravalvular aortic

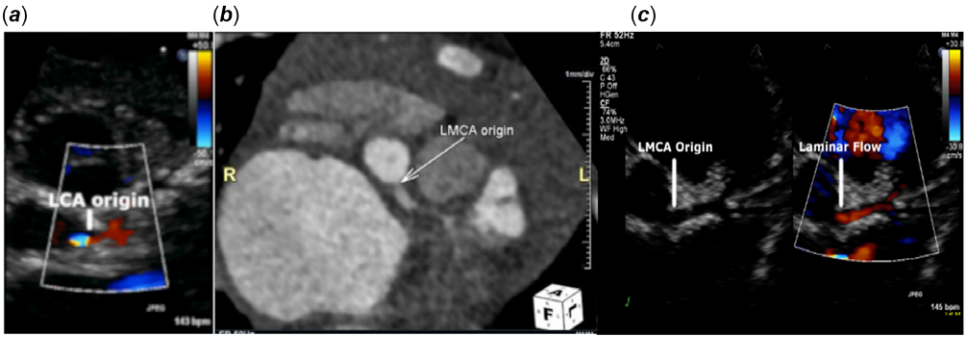

Figure 1. (***a***) Echocardiogram in the parasternal axis view showing color Doppler aliasing in the left coronary artery origin. (***b***) Computed tomographic angiogram showing narrowing of the left main coronary origin. (***c***) This post-operative image from a PSAX view demonstrates a widely patent left coronary artery ostium with laminar flow by color Doppler.

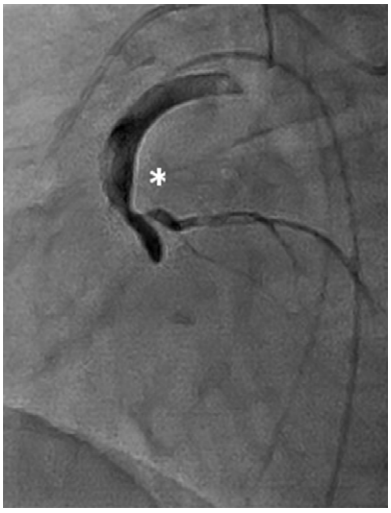

Figure 2. Angiogram obtained during cardiac catheterisation. *Indicates left coronary artery origin with narrowing.

stenosis.[2–5] Isolated left coronary ostial stenosis and atresia are more rare.[6–8] Both can present similarly with failure to thrive, angina that presents as irritability with feeding, and ischaemic cardiomyopathy in infancy, though multiple cases of initial diagnosis in older patients have been reported.

In an infant presenting with cardiomyopathy and coronaries arising from their appropriate sinuses, left main coronary ostial stenosis is an important diagnosis to consider. It requires a high index of suspicion and careful echocardiographic analysis. In this case, a second review of the initial echocardiogram was important in the initiation of further evaluation and ultimately, treatment. This strategy should be carefully considered when the clinical picture does not match findings from the echocardiogram. Laboratory and electrocardiogram findings are useful to increase the index of suspicion for coronary disease in these patients. Multimodality imaging with non-invasive computed tomographic angiography and invasive angiography should be considered. Prompt diagnosis is important as ischaemia is ongoing and the chance for myocardial recovery decreases as the length and severity of ischaemia increase.

Even when it is an isolated lesion, surgical revascularisation is indicated. This case demonstrates that successful repair of this lesion can lead to the return of normal cardiac function. There is no standard of care as there are so few reported cases of left main coronary ostial stenosis as an isolated lesion. The pericardial patch has proven to be an adequate means of repair, demonstrated by the fact that the patient's left ventricular ejection fraction has remained stable over 4 years of follow-up. Other cases have been reported of patients presenting in early childhood with severe congenital obstruction of the left main coronary ostium. Three cases were repaired using an azygos vein graft or patch.[10] The success in our case of the pericardial patch supports the use of direct ostial repair rather than coronary artery bypass grafting.

Supplementary material. To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951120003224

Acknowledgements. None.

Financial support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of interest. one.

Ethical standards. Not Applicable.

References

1. Xiao Y, Jin M, Han L, et al. Two congenital coronary abnormalities affecting heart function: anomalous origin of the left coronary artery from the pulmonary artery and congenital left main coronary artery atresia. Chin Med J (Engl) 2014; 127: 3724–3731.
2. Goel P, Madhu Sankar N, Rajan S, Cherian KM. Coarctation of the aorta, aortic valvar stenosis, and supravalvar aortic stenosis with left coronary artery ostial stenosis: management using a staged hybrid approach. Pediatr Cardiol 2001; 22: 83–84.
3. Satran A, Dawn B, Leesar MA. Congenital ostial left main coronary artery stenosis associated with a bicuspid aortic valve in a young woman. J Invasive Cardiol 2006; 18: E114–E116.
4. Yildiz O, Altin FH, Kaya M, Ozyılmaz I, Guzeltas A, Erek E. Left coronary artery stenosis causing left ventricular dysfunction in two children with supravalvular aortic stenosis. World J Pediatr Congenit Heart Surg 2015; 6: 311–316.
5. Pieles GE, Ofoe V, Morgan GJ. Severe left main coronary artery stenosis with abnormal branching pattern in a patient with mild supravalvar aortic stenosis and Williams-Beuren syndrome. Congenit Heart Dis 2014; 9: E85–E89.
6. Basso C, Frescura C, Corrado D, et al. Congenital heart disease and sudden death in the young. Hum Pathol 1995; 26: 1065–1072.
7. Fabozzo A, Vida VL, Reffo E, et al. Intraoperative diagnosis of bilateral coronary ostia stenosis: a rare case of ischemic heart disease in a 3-month-old patient. Ann Thorac Surg 2011; 92: 1875–1877.
8. Laux D, Bessières B, Houyel L, et al. Early neonatal death and congenital left coronary abnormalities: ostial atresia, stenosis and anomalous aortic origin. Arch Cardiovasc Dis 2013; 106: 202–208.
9. Durán AC, Arqué JM, Sans-Coma V, Fernández B, de Vega NG. Severe congenital stenosis of the left coronary artery ostium and its possible pathogenesis according to current knowledge on coronary artery development. Cardiovasc Pathol 1998; 7: 261–266.
10. Tominaga T, Asou T, Takeda Y, et al. Coronary angioplasty for congenital obstruction of the left main coronary artery. Gen Thorac Cardiovasc Surg 2016; 64: 156–159.