

Rare Congenital Obstruction of the Inferior Vena Cava Diagnosed by Intraoperative Transesophageal Echocardiography



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INTRODUCTION

We present a case of rare congenital obstruction of the inferior vena cava (IVC) outlet detected intraoperatively on transesophageal echocardiography (TEE). Lacking a better explanation, hepatic dysfunction had been attributed to primary presentation of hypertrophic cardiomyopathy with presumed diastolic heart failure. However, the intraoperative finding of IVC obstruction better explained hepatic disease, and concurrent repair was successful in resolving the hepatic dysfunction.

CASE PRESENTATION

A 40-year-old patient with progressive symptoms of hypertrophic cardiomyopathy was referred for septal myectomy. His only presenting symptom was dyspnea with exertion. There were no signs of portal hypertension. There were no other signs of left- or right-sided heart failure, including absence of pulmonary edema and jugular venous pulse elevation. His preoperative evaluation laboratory results were notable for mild transaminitis with aspartate transaminase of 70 U/L and alanine transaminase of 92 U/L. Total bilirubin and international normalized ratio were normal.

Preoperative transthoracic echocardiography (TTE), although it predated the operative period by many months, demonstrated a resting left ventricular outflow tract gradient of 100 mm Hg, moderate diastolic dysfunction, and mild to moderate mitral regurgitation. Subcostal views demonstrated an IVC of 2.1 cm. Although on close retrospective examination, the atriocaval junction could appear narrowed (Video 1), this may also be attributed to foreshortening as it comes in and out of view with each beat. Similarly, although the color Doppler in this view (Video 2) may have a trace jet, it may be missed unless focused on. In either case, the official read commented on neither.

A hepatology consult was obtained and ultrasound performed, without evidence of cirrhosis. The patient had no risk factors for infectious hepatitis, so hepatology did not think that waiting for serologic results was necessary in the absence of impaired function. The leading

differential diagnosis was congestive hepatopathy from diastolic dysfunction or mitral regurgitation or drug-induced hepatitis from verapamil. The recommendation was to proceed with surgery with postoperative hepatologic follow-up.

After induction of general anesthesia, baseline TEE confirmed preoperative echocardiographic findings, with the exception of the mitral regurgitation, which was mild. However, in the midesophageal bicaval view, color flow Doppler interrogation revealed turbulent flow emanating from the IVC (Video 3). The jet originated at the level of the Eustachian valve and had a measured orifice width of 0.8 cm (Figure 1), approximately half the width of the preceding cava. After an uneventful septal myectomy, the right atrium was exposed to reveal a constricted, nonseptated Eustachian valve (Figure 2). A strip of tissue approximately 0.5 × 2 cm (Figure 3) was excised, thereby opening the atriocaval junction completely. At the conclusion of the procedure, TEE confirmed unrestricted flow from the IVC (Video 4). On the basis of these findings, we surmised that the flow restriction accounted for the congestive hepatopathy, and indeed 5 weeks later at a follow-up visit, the patient's transaminase levels had returned to normal.

DISCUSSION

Idiopathic IVC obstructions are extremely rare by report.¹⁻³ Symptoms present similarly to acquired obstructions such as Budd-Chiari syndrome and can include lower body venous congestion, hepatic congestion with associated hepatitis, portal hypertension, edema, and ascites.¹ Lesions can be the result of aberrant membranes that can form either in the vena cava itself or the hepatic veins, vessel stenosis, or malformed Eustachian valve.^{2,4} More severe cases present early in childhood and carry high mortality unless detected and repaired.⁴

In our patient, the restriction was small enough to be subclinical, though it may have progressed over time with the sequela of chronic hepatic congestion.⁵ There are no data in the literature regarding the natural history of progression of this presentation. His concurrent cardiac problems led to early diagnosis through preoperative suspicion and intraoperative imaging. Conventional preoperative TTE did not detect an obstruction, which is not unexpected. Although TTE can provide some visualization of the IVC outlet and Eustachian valve in the views listed in Table 1,⁶ it may not be sufficiently sensitive, as some views are not part of a standard examination, others may incompletely visualize the IVC and Eustachian valve together,^{6,7} and all may be limited by poor windows or habitus.

TEE, however, does enable a good view of the IVC outlet in the midesophageal bicaval view, which is part of the standard perioperative examination.⁸ More of the IVC can also be exposed by advancing the probe from this view to include the hepatic veins, although this is not a standard view (Table 2). The Eustachian valve is a normal

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VIDEO HIGHLIGHTS

Video 1: Preoperative TTE, subcostal long-axis IVC view with possible narrowing of atriocaval junction (retrospective read).

Video 2: Preoperative TTE, subcostal long-axis IVC view with possible trace jet apparent on color Doppler (retrospective read).

Video 3: Intraoperative TEE, baseline, midesophageal bicaval view with prominent jet on color Doppler.

Video 4: Intraoperative TEE, closure, midesophageal bicaval view with resolution of flow restriction on color Doppler.

View the video content online at www.cvcasejournal.com.

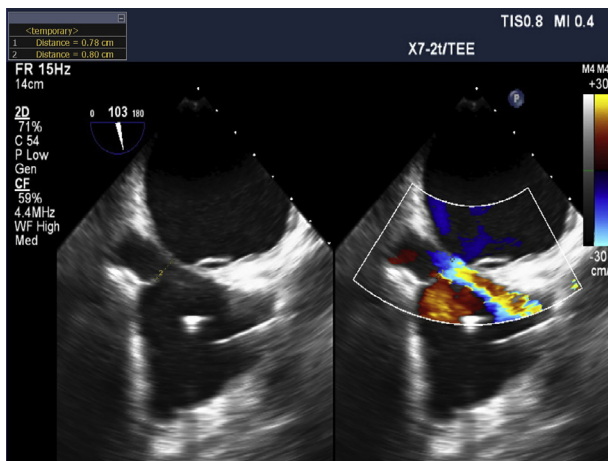


Figure 1 Intraoperative TEE, baseline, measurement of restriction orifice width.

echocardiographic finding and can be prominent without being obstructive because of its usual septation.^{2,9} Obstruction would be visible with color Doppler, although gradients may be difficult to assess because of orthogonal views. Three-dimensional imaging may help assess the morphology of the obstructive tissue, although we did not pursue this at the time, because of time constraints. Severity of obstruction and need for intervention would depend then on symptomatology, but we could find no literature on natural history or specific guidelines on need, timing, or modality of intervention. That said, all references to repair we could find were surgical in nature. Although additional preoperative testing (e.g., venography or TEE) in retrospect may have elucidated the etiology, we would posit that this condition is sufficiently rare that the pretest probability remains too low to risk complications of routine invasive testing on the basis of the clinical presentation alone.

In our patient, as previously commented, the images were not sufficiently distinct, and the subtle changes, clearer in hindsight, were not noted. These observations admittedly may be confounded by differing volume status at different times as well as the patient's being under anesthesia. In the end, preoperative workup, whether because of technical limitations or human error, is not always 100% sensitive.

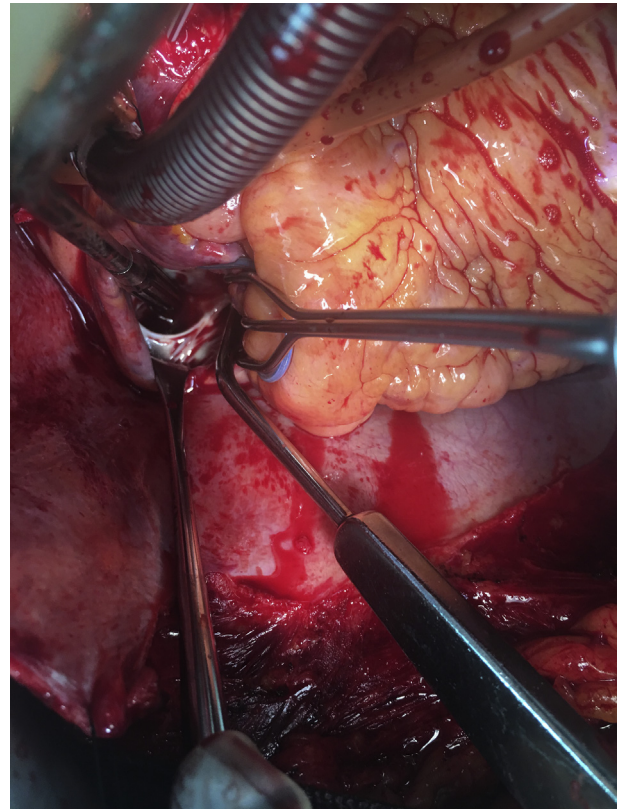


Figure 2 Obstructive membrane in situ. The IVC outlet with constricting membrane is visible through the atriotomy.



Figure 3 Excised membrane, 0.5 × 2 cm excised tissue from IVC outlet.

Table 1 Views including IVC and Eustachian valve on TTE

1. Subcostal long-axis IVC*
2. Apical four-chamber, posterior angulation (coronary sinus view)
3. Parasternal long-axis, RV inflow view
4. Subcostal SVC (bicaval) view

RV, Right ventricular; SVC, superior vena cava.

*Standard examination view.

Table 2 Views including IVC and Eustachian valve on TEE

1. Midesophageal bicaval view*
2. Midesophageal bicaval view: deep, including hepatic veins

*Standard examination view.

Intraoperative TEE can be a final opportunity to detect overlooked or subclinical pathology.

CONCLUSION

We demonstrated a rare idiopathic finding on intraoperative TEE amenable to immediate repair that was not clearly evident on preoperative TTE. In this case, TEE may have been more sensitive than TTE for detecting this rare problem. True of all imaging modalities, and especially in a high-productivity environment, it can be tempting to perform more focused examinations for the anticipated problems. Even on complete examinations, subtle findings can be missed unless prominent in optimal views, especially when unanticipated. This case underscores the value of complete and systematic intraoperative TEE as part of the standard intraoperative anesthetic management for cardiac surgery.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.01.003>.

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