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Case Report

The Role of Intraoperative Transesophageal Echocardiography in Identifying a Fenestrated Occlusion of the Inferior Vena Cava During Pulmonary Thromboendarterectomy

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ANTIPHOSPHOLIPID SYNDROME (APLS), both primary and secondary, is a known cause of recurrent thrombotic events, leading to deep vein thrombosis and pulmonary embolism. Chronic thromboembolic pulmonary hypertension (CTEPH) is known to result from recurrent episodes of pulmonary embolism and is the main indication for pulmonary thromboendarterectomy (PTE) surgery, a procedure that significantly improves symptoms and the prognosis of these patients. ²

Membranous obstruction of the inferior vena cava (MOVC) is a recognized clinical entity of unclear origin, being either congenital or acquired, and is the most common cause of Budd-Chiari syndrome worldwide.³ It can be associated with systemic disorders such as systemic lupus erythematosus (SLE) (1%) and thrombophilic conditions such as APLS (0%-17%). However, an etiologic factor cannot be identified in up to 30% of these patients. Presentation can be acute,

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causing hepatic failure, or more commonly chronic, eventually leading to cirrhosis, ascites, hepatomegaly, and portal hypertension if left untreated.

The authors present a case in which the use of intraoperative transesophageal echocardiography (TEE) before bypass during PTE surgery was used to identify a membranous obstruction of the inferior vena cava-right atrium (IVC-RA) junction in a patient with a history of SLE and secondary APLS. This altered surgical management with the addition of open resection of the membrane, highlighting the importance of TEE in guiding the surgical procedure.

Case Presentation

A 26-year-old female with chronic thromboembolic pulmonary disease associated with SLE and APLS presented to the authors' department for PTE surgery with cardiopulmonary bypass (CPB) and deep hypothermic circulatory arrest.

The patient reported severe, progressively worsening shortness of breath (World Health Organization functional class III). In the past, she experienced multiple clinical episodes of deep vein thrombosis and pulmonary embolism,

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the last episode taking place 4 years ago. Her medical history included SLE, APLS, and Raynaud's syndrome. The patient was taking regular steroids, immunosuppressive drugs (azathioprine), sildenafil, hydroxychloroquine, and anticoagulants (warfarin, bridged to low-molecular-weight heparin before the surgery).

The preoperative transthoracic echocardiogram showed good biventricular function. According to the right heart catheterization, hemodynamic parameters were as follows: cardiac index 3.4 L/min/m², systolic pulmonary artery pressure (PAP) 41 mmHg, diastolic PAP 19 mmHg, mean PAP 29 mmHg, and pulmonary vascular resistance 200 dyn/s/cm⁵. A magnetic resonance angiography scan showed dilation of the main pulmonary artery (2.8 cm diameter), cardiac chambers of normal size, occlusion of the right lower lobe pulmonary artery, and severe attenuation of the right middle lobe branches. The decision to proceed to the PTE procedure was made on the basis of deteriorating clinical status.

In addition to the pulmonary findings, 2 chest and abdominal computed tomography scans from 2014 and 2015 showed common iliac vein and IVC obstruction reaching the confluence of the hepatic veins. In addition, there was a distended azygos vein and an extended network of collateral veins draining to the superior vena cava (SVC), indicating longstanding obstruction rather than congenital absence of the IVC. Also, the liver was congested, possibly due to hepatic vein-IVC confluence occlusion. However, the patient did not demonstrate any clinical or laboratory signs of hepatic impairment. In view of the aforementioned findings, the surgical plan was changed, and it was decided not to use standard bicaval cannulation for CPB. Instead, a cannula was placed into the SVC as usual, but a Ross basket was used to provide drainage from the RA, with a plan to provide additional venous drainage via the femoral venous route if necessary.

Induction of general anesthesia proceeded uneventfully after application of standard monitoring and placement of a peripheral intravenous and radial arterial line. A 4-lumen central venous line and a pulmonary artery catheter were placed in the right jugular vein under ultrasound guidance and

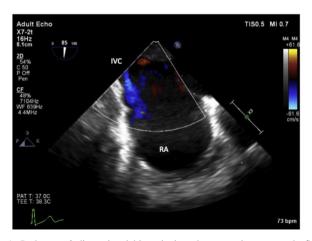


Fig 1. Prebypass 2-dimensional bicaval view demonstrating antegrade flow from the IVC to the RA. IVC, inferior vena cava; RA, right atrium.

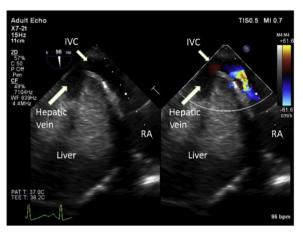


Fig 2. Prebypass 2-dimensional color IVC view. Extension of the bicaval view to the IVC showing turbulent flow arising just distal to IVC-RA junction. IVC, inferior vena cava; RA, right atrium.

a second arterial line was inserted in the patient's right femoral artery as per normal practice. TEE examination before bypass showed good biventricular function overall, with only mild impairment of the right ventricle (RV) (Fractional area change 33%, mildly impaired movement of the right ventricular free wall at apical level), and normal size atria without any signs of a patent foramen ovale. A clot was detected in the right pulmonary artery, whereas the left pulmonary artery could not be visualized. On the bicaval view, antegrade flow was detected from the IVC into the RA (Fig 1), and when the view was extended toward the IVC, a fenestrated, membranelike structure was detected just distal to the junction between the IVC and the RA. The membrane caused partial occlusion of flow, and 2 jets of accelerated, turbulent flow could be visualized (with color-flow Doppler) (Figs 2 and 3; Videos 1 and 2). Examination of the hepatic veins with color-flow Doppler showed normal, antegrade flow.

The surgeon was notified and with confirmation of the partial obstruction to the opening of the IVC at the cavo-atrial junction, the modified cannulation strategy previously described was undertaken successfully. CPB was instituted

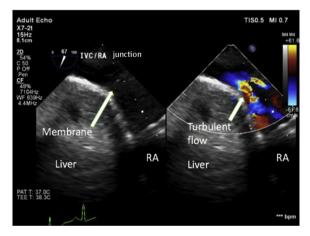


Fig 3. Prebypass 2-dimensional color view of the IVC-RA junction depicting the existence of a membrane just distal to the junction causing partial obstruction and turbulent flow. IVC, inferior vena cava; RA, right atrium.

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