Title:

Syncope and pregnancy management in patient with Large Coronary Artery Fistula and Right Atrial Varix

Authors: Garagiola Maria Luz, Smettei Osama, Blissett Sarah

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- Clinical Presentation:

An 18-year-old female with a history of recurrent intermittent dizziness and palpitations presented to the emergency department after experiencing palpitations followed by syncope, occurring in the context of a gastrointestinal viral infection. Transthoracic echocardiography revealed an unknown large, mobile, serpiginous, irregular structure with calcified rims in the right atrium, prompting admission and further investigation. Transesophageal echocardiography, right and left heart catheterization, cardiac computed tomography, and myocardial perfusion imaging were completed. These investigations identified a dilated circumflex with fistulous communication to the coronary sinus (17 × 26mm) terminating in a 35mm exophytic varix within the right atrium (Figure 1).

The patient was referred for a surgical consultation but became pregnant before completing the assessment. Given the complexity of her new diagnosis, she was closely monitored through a specialized Cardio-Obstetrics program, including serial echocardiograms to monitor any pregnancy-related complications. These assessments played a crucial role in guiding the management of both her pregnancy and delivery. During this time, she experienced no significant complications.

- Imaging Findings:

Transthoracic and transesophageal echocardiography confirmed that the varix did not obstruct the tricuspid valve inflow, ruling out valvular obstruction as a contributing factor to her symptoms. High velocity continuous flow (peak systolic gradient of 76mmHg) from the coronary sinus varix to the right atrium was identified (Figure 1-F). This finding suggests the possibility of increased blood flow in a low preload state, which potentially could increase flow from the coronary bed to the fistulous tract. Heart catheterization demonstrated normal left and right-sided filling pressures and cardiac index, without hemodynamically significant shunt (Qp:Qs 1.3:1). Holter monitoring demonstrated her rhythm was in sinus when she felt palpitations and dizziness.

- Summary/Discussion Points:

Coronary arteriovenous fistulas are abnormal conduits between a coronary artery and another cardiovascular lumen, bypassing the capillary bed. The reported prevalence is <0.1%. Potential physiologic consequences of this case include rupture, coronary steal, thrombotic risk, heart failure, and potential tricuspid valve obstruction.

This case underscores the critical role of echocardiography in evaluating syncope, the utility of multimodality imaging in diagnosing complex structural heart disease, and the importance of longitudinal echocardiographic monitoring in high-risk pregnancies.

Figure 1:

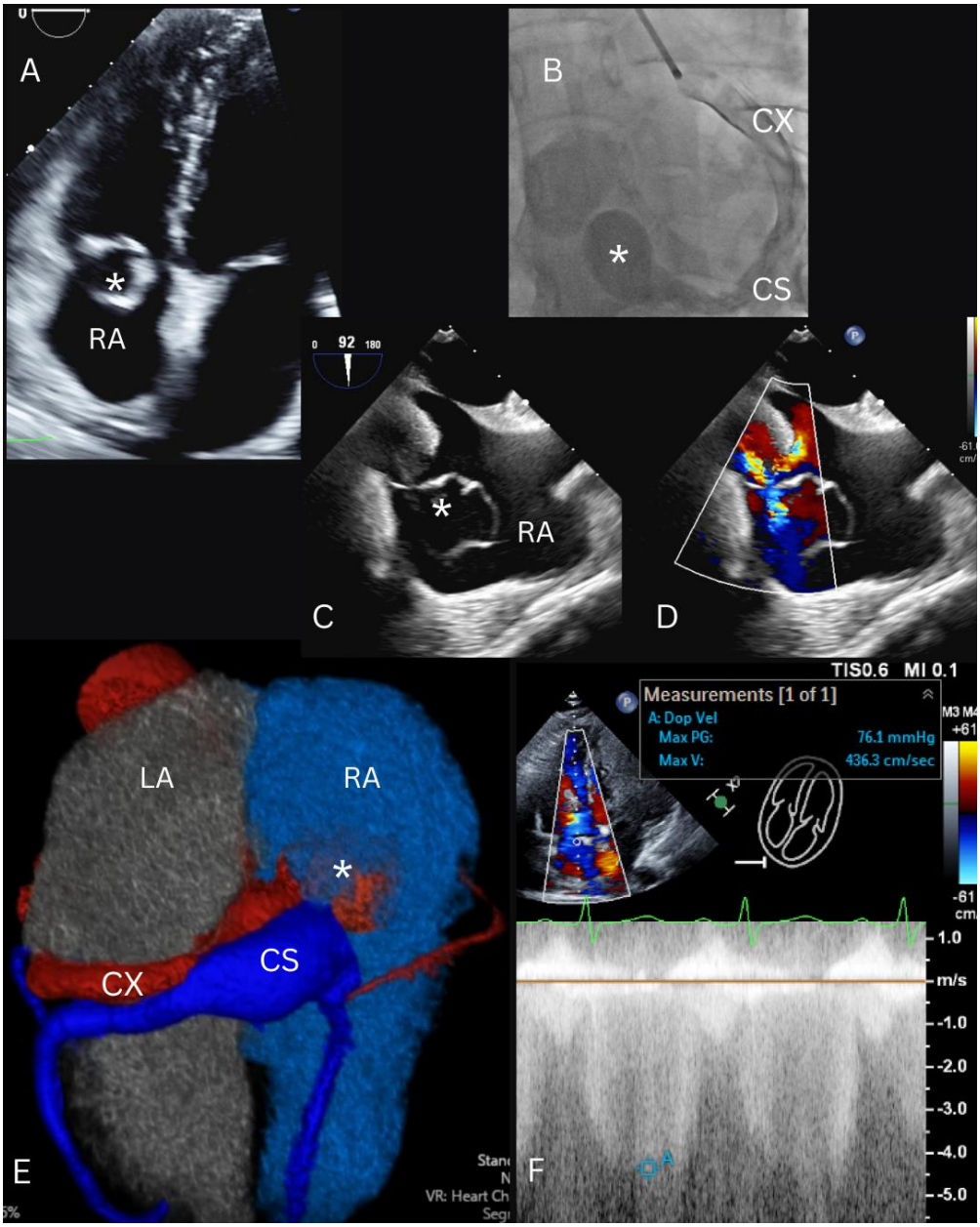

Figure 1: Large circumflex artery fistula to coronary sinus terminating in an exophytic varix. A: Transthoracic echocardiogram showing exophytic varix at the right atrium in close proximity to the tricuspid valve; B: Coronary angiography demonstrating the large circumflex to coronary sinus fistula, exophytic varix, and flow from the exophytic varix to the right atrium; C and D: Transesophageal echocardiography with color Doppler demonstrating flow into the right atrium and the large exophytic varix; E: Rendering reconstruction of cardiac computed tomography with emergency of the aneurysmatic left circumflex connected to coronary sinus which terminates in an exophytic varix variant in the right atrium; F: Doppler interrogation of peak flow from coronary sinus varix to right atrium in transthoracic echocardiogram, maximum gradient 76mmHg. *: exophytic varix, CS: coronary sinus, CX: circumflex artery, LA: left atrium, RA: right atrium.