Percutaneous Closure of Aorto-Right Atrial Fistula: An Interesting Case of Marfan's Syndrome and Fistula Formation

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Case Presentation

A 43-year-old male with a history of Marfan's syndrome, coronary artery disease and mitral valve endocarditis status-post mitral valve annuloplasty was admitted with NYHA class IV symptoms and cardiogenic shock. Two distinct color-flow jets on transesophageal echocardiography (TEE) supported evidence for fistulae formation at the aortic sinus and the right atrium (RA). After stabilization, closure options were discussed with the patient and a multi-disciplinary team including a cardiothoracic surgeon, structural heart disease specialist and heart failure specialist. Shared decision for percutaneous closure of the fistula was made. Figure 1A, demonstrates a peri-operative TEE with two distinct fistulae with color-flow jets emanating from the sinus of Valsalva into the RA. Each fistula orifice measured approximately 3mm in diameter with 5mm of separation. Under fluoroscopy, trans-aortic wire and sheath were used to cross a single fistula into the RA. An 18mm occluder device (Figure 1B and 1C), was deployed via transaortic into the RA. Intracardiac echocardiography revealed good device position and trivial jet. Post-operative TEE verified successful occlusion of both fistulae. The patient tolerated the procedure and was discharged. He was seen in clinic two weeks post-discharge and reported resolution of his dyspnea.

Conclusion

Aorto-atrial fistulas (AAF) are rare, complex and life-threatening; however, potentially treatable conditions. While most cases are acquired, congenital causes exist. This is an interesting case that illustrates a unique presentation of a rare condition and showcases potential for endovascular treatment. There are no randomized clinical trials and more scientific evidence is required to establish recommendations for a diagnostic and treatment algorithm. Until then clinical judgment with consideration for percutaneous closure seems a viable option.

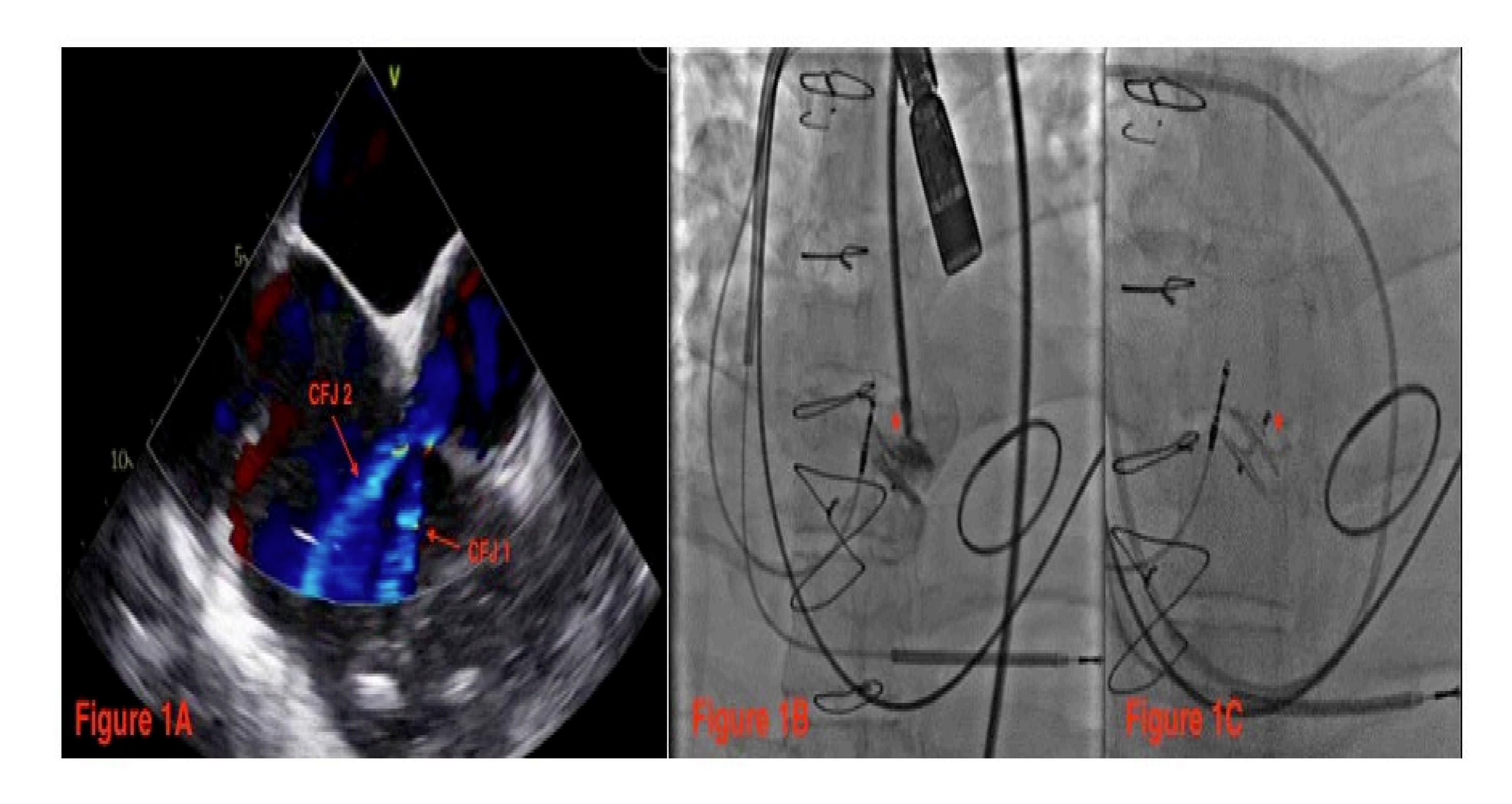


Figure 1A demonstrates a peri-operative transesophageal echocardiogram (TEE) with two distinct fistulae with color flow jets (CFJ1 and CFJ2) Figure 1B and Figure 1C demonstrate the successful closure of the fistulae with a closure device.

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