The missing leak: a case report of a baffle-leak closure using real-time 3D transoesophageal guidance

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Brady-arrhythmias in patients undergone atrial switch procedures (Mustard or Senning procedure) for complete transposition of the great arteries (TGA) are common and often require implantation of permanent pacemakers. It has been shown that in patients with palliated congenital cardiac defects with residual intra-cardiac shunts, permanent pacemaker implantation is associated with an increased risk of thrombo-embolism. Patients with TGA and concomitant baffle leaks may have an even further increased thrombo-embolic risk, given that the leaks can provide the conduit for venous to systemic thromboembolism. In order to decrease this risk, all TGA patients who require pacemaker implantation typically undergo a thorough pre-procedural evaluation to assess for the presence of a baffle leak. Traditional imaging modalities, however, are often limited in their ability to detect and/or properly locate small baffle leaks. We report a case of a patient with TGA and a baffle leak that was both identified and percutaneously closed with the assistance of real-time 3D transoesophageal echocardiography.

Introduction

One of the most common long-term complications of the atrial switch procedure for complete transposition of the great arteries (TGA) is the development of brady-arrhythmias and/or sinus node dysfunction often necessitating implantation of a permanent pacemaker.1–5 Transvenous pacemaker leads have been shown to double the risk of thrombo-embolism in patients with intracardiac shunts,6 including deliberately created interatrial communications and baffle leaks.7,8 The exact mechanism behind this increased risk is currently unknown. Regardless, given this elevated thrombo-embolic risk, it is imperative that these patients undergo a thorough diagnostic evaluation prior to pacemaker implantation to specifically exclude baffle leaks and then subsequent closure of these leaks.9

Baffle leaks can be identified using angiography, echocardiography, magnetic resonance imaging (MRI), and computed tomography angiography (CTA).10–12 Small baffle leaks are more common than obstruction and often can be visualized only by transoesophageal echocardiography (TEE).12 We present a case of a young woman with TGA who required a permanent pacemaker for sinus node dysfunction. Baffle evaluation using saline contrast transthoracic echocardiography (C-TTE), 2D TEE, and MRI suggested a baffle leak, but failed to identify the exact anatomical location of the leak. Angiography in multiple planes actually failed to identify any leak at all. Subsequent interrogation using real-time 3D transoesophageal echocardiography (RT3D TEE), however, revealed a small leak on the superior border of the superior baffle limb that was percutaneously closed using RT3D TEE guidance. This is the first reported case, to our knowledge, using RT3D TEE to both diagnose as well as guide a baffle leak closure.

Case report

A 31-year-old female who had undergone a Mustard procedure at the age of 20 months for TGA was seen in our Adult Congenital Heart Disease clinic for evaluation prior to becoming pregnant. Cardiopulmonary exercise stress testing revealed a peak VO2 of 23 mL/kg per minute and a normal chronotropic response to exercise. During recovery phase, however, the patient developed a junctional bradycardia that was associated with light headedness and marked hypotension consistent with vasodepressor near-syncpe. A Holter monitor was then placed, which revealed long periods of junctional escape rhythm. Given the haemodynamic demands of pregnancy, there was a concern that there was enough evidence of sinus node dysfunction, and that a permanent pacemaker was recommended. Prior to...
proceeding with pacemaker implantation, given the risk of pacemaker lead infection and paradoxical emboli if a baffle leak is present, a transthoracic echocardiogram with agitated saline contrast was performed. This study suggested that there was a small baffle leak though it could not be located on transthoracic echocardiography. Given these findings, a cardiac MRI was performed to evaluate for the presence of a baffle leak and more accurately assess right ventricular systolic function. The cardiac MRI revealed patent superior and inferior baffles with no evidence of a baffle stenosis. Volumetric and flow analysis demonstrated left (pulmonary venous) to right (systemic venous) shunting with a main pulmonary arterial flow of 43.1 mL/m$^2$ and an ascending aortic flow of 34.8 mL/m$^2$, which is a calculated Qp:Qs of 1.2. On cine imaging, however, a discrete baffle leak could not be identified. Careful inspection of both baffles, however, failed to reveal the source of this increased flow. In light of this finding, the patient was subsequently referred for cardiac catheterization with concomitant RT3D TEE to definitively diagnose or exclude a baffle leak.

A 5 French (Fr) pigtail catheter was advanced from the right femoral vein into the inferior limb of the baffle. Baffle angiography in a straight AP and lateral projections revealed no evidence of obstruction or leak (Figure 1A and B). A 7Fr NIH Cardiomarker catheter (Medtronic, Minneapolis, MN, USA) was then advanced from the right internal jugular vein into the superior limb of the baffle and subsequent angiography in multiple planes again revealed no evidence of obstruction or leak (Figure 1C and D).

Intraprocedurally TEE was performed using an X7-2t TEE transducer and iE33 ultrasound system (Philips Medical Systems, Andover, MA, USA). After administration of agitated saline contrast, bubbles were seen on the pulmonary venous side of the baffle consistent with a small baffle leak. Careful inspection of both limbs of the baffle by the 2D TEE mode, failed to reveal the exact location of the leak. Despite angiographic absence of a baffle leak, careful inspection of the superior limb of the baffle using RT3D TEE directed us to a small area of turbulence on Colour flow Doppler (2D TEE) at the superior border of the superior limb of the baffle consistent with a small baffle leak (Figure 2A and B). Review of the angiographic images and 2D TEE images again failed to reveal the precise location of the leak necessitating RT3D TEE guidance to assist in defect closure.

A JR4 catheter was then advanced over a 0.035 in. Glide wire (Terumo Corp. Piscataway, NJ, USA) from the right internal jugular vein to the site of the leak and, although the Glidewire and JR4 catheter were successfully manipulated across the defect into the pulmonary venous side of the baffle, the tortuous course navigating to the baffle leak caused the JR4 catheter to repeatedly prolapsed back from the pulmonary venous to the systemic venous side of the baffle. The orientation of flow seen by TEE predicted a more direct access to the leak coming from an inferior approach given the leak’s superior location. The baffle leak was then subsequently easily crossed from the femoral approach using RT3D TEE guidance and the Glidewire and JR4 catheter were advanced into the left upper

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**Figure 1** Baffle angiography: (A and B) inferior limb baffle angiography in the anterior–posterior and lateral projections demonstrating no angiographic evidence of a leak with only minimal narrowing. (C and D) Superior limb baffle angiography in the anterior–posterior and lateral projections demonstrating no angiographic evidence of a leak (LV, morphologic left ventricle, functioning as the subpulmonary ventricle).
The real-time 3D orientation allowed easy direction of the catheter and system across the baffle leak from the femoral vein within 2–3 min.

Evaluation of the baffle leak using RT3D TEE revealed a 2–3 mm diameter leak, and the decision was made to use a 4 mm Amplatzer Septal Occluder (ASO) device (AGA Medical, Golden Valley, MN, USA) for defect closure. The Glidewire was exchanged for an Amplatz 6 cm soft-tip super stiff wire (Boston Scientific, Natick, MA, USA) over which a 7Fr Amplatzer TorqVue delivery system (AGA Medical) was advanced. The 4 mm Amplatzer ASO was then successfully deployed in the standard fashion as described previously.13,14 The entire procedure was performed using RT3D TEE guidance (Figure 3A–F).

Post-procedural echocardiographic inspection of the device confirmed the absence of a residual shunt.

Following her percutaneous baffle leak closure, the patient underwent successful permanent pacemaker implantation without complication and currently is in her third trimester of pregnancy.

Discussion

This definitive case report illustrates the novel use of RT3D TEE in the diagnosis and treatment of a baffle leak. Importantly, the case highlights the limitations of angiography, MRI and, surprisingly, 2D TEE in detecting and assisting in the closure of a small baffle leak and demonstrates the utility of RT3D TEE guidance in percutaneous baffle leak closure.

Patients with TGA who have undergone surgical palliation with an atrial switch procedure and who subsequently develop the need for a pacemaker represent a challenging population. Thorough assessments of both baffle limbs for the presence of small leaks that may pose an increased thrombo-embolic risk is often difficult when using traditional invasive and non-invasive imaging modalities. In our case, though the patient’s superior baffle leak was suggested by the traditional imaging techniques, its exact anatomical location was only revealed when RT3D TEE was used. This modality not only provided the diagnosis but also assisted in better understanding the 3D geometry and location of the defect to allow for more efficient percutaneous closure of the defect. Although small, this hole was in close proximity to were the atrial pacing lead was placed, and hence its location may have facilitated even a small thrombus on the lead to embolize systemically.

Our case is the first known case using RT3D TEE in the assessment and percutaneous closure of a baffle leak in an
adult patient with TGA. Because of its unique ability to define both spatial relationships and anatomic anomalies in real time and with remarkable clarity, RT3D TEE represents a new, unique, and exciting imaging modality that is quickly becoming an integral part of our interventional structural heart disease practice. Only recently has the availability of RT3D TEE extended to beyond select research centres, and as this case report illustrates, our understanding of its potential applications in structural heart disease continues to be a work in progress. As clinical experience with RT3D TEE continues to grow, however, so too will the interventional cardiology community’s understanding of its value in all realms of cardiovascular disease.

References


