Tricuspid Valvular Heart Disease:
The Forgotten Valve

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As the primary focus of daily clinical practice involves pathologies of the left side of the heart, including aortic and mitral valvular disease, abnormalities of the tricuspid valve (TV) are often overlooked.[1,2] The resulting functional impairments from TV abnormalities often lead to increased patient morbidity and mortality, underlining the importance for early recognition of this valvular disease.[1] ECHO is an effective non-invasive diagnostic imaging modality for characterizing TV disorders. Prior to patient referral for ECHO, it is necessary for physicians to be aware of a simple classification of TV disease, including both primary and secondary etiologies.

Primary TV disease is associated with intrinsic valvular pathology, while secondary TV disease arises from annular dilation secondary to right ventricular (RV) dilation and dysfunction. Primary TV disorders include infective endocarditis (IE), Ebstein's anomaly, rheumatic heart disease, carcinoid heart disease and TV papillary muscle rupture. Secondary TV disease involves disorders of the RV leading to annular dilatation including primary pulmonary hypertension (PPH), secondary pulmonary hypertension and arrhythmogenic RV dysplasia (ARVD). The following case scenarios focus on representative examples of both primary and secondary TV disease.

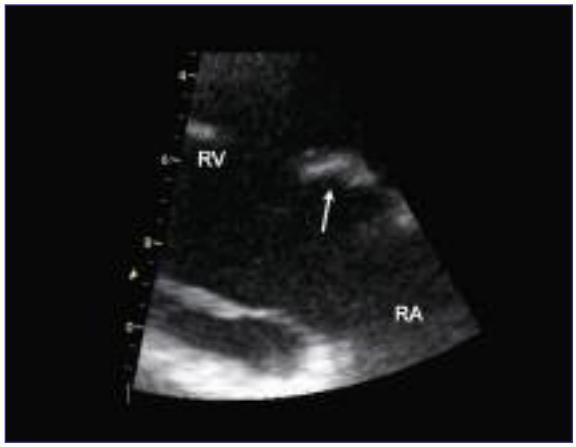

Figure 1. A right ventricular (RV) inflow view on transthoracic ECHO demonstrating an echodense mass attached to the anterior leaflet of the tricuspid valve (TV) (arrow) consistent with a vegetation. Right atrium (RA).

IE

An 18-year-old college student presented with acute shortness of breath (SOB). On physical examination, the BP was 100/60 mmHg, sinus tachycardia of 102 bpm and febrile at 38.5˚C. The jugular venous pressure (JVP) was elevated at 6 cm above the sternal angle, with a grade IV/VI pansystolic murmur heard prominently in the lower left sternal border (LLSB) that increased upon inspiration. ECHO confirmed an echodense mass attached to the anterior leaflet of the TV, consistent with a vegetation (Figure 1).

IE of the TV is predominant amongst select populations including IV drug users and patients with indwelling venous catheters.[1-3] A commonly associated complication is septic pulmonary emboli due to the increased mobility of the mobile vegetations attached to the TV leaflets.[3] Treatment varies based on the severity of the infection, commonly including a combination of antimicrobial therapy, surgical repair of TV and/or TV replacement as in this case.

Ebstein's anomaly

A 32-year-old male postal worker presented with dyspnea on exertion, cyanosis, palpitations and two separate transient ischemic attack (TIA) events in the past six months. Physical examination findings revealed a BP of 130/70 mmHg, normal sinus rate of 72 bpm, an elevated JVP with prominent V wave and a pansystolic murmur at LLSB that increased with inspiration. ECHO confirmed apical displacement of the septal leaflet of the TV as compared to the anterior leaflet of the mitral valve with "atrialization" of the RV (Figure 2). The concomitant presence of an interatrial shunt was consistent with the diagnosis of Ebstein's anomaly.

Ebstein's anomaly originates from a congenital malformation of the TV and occurs in < 1% of patients with congenital heart disease (CHD).[4] Associated characteristics include apical displacement of the septal tricuspid leaflet > 11 mm as compared to the mitral valve leaflet, "atrialization" of the RV, RV dysfunction and the presence of an interatrial shunt in 25% of patients.[5] Approximately 25% of patients with Ebstein's will demonstrate an accessory Wolff-Parkinson-White pathway. Patients with resting cyanosis, New York Heart Association Class III or above or incessant right-sided heart failure are candidates for surgical repair.

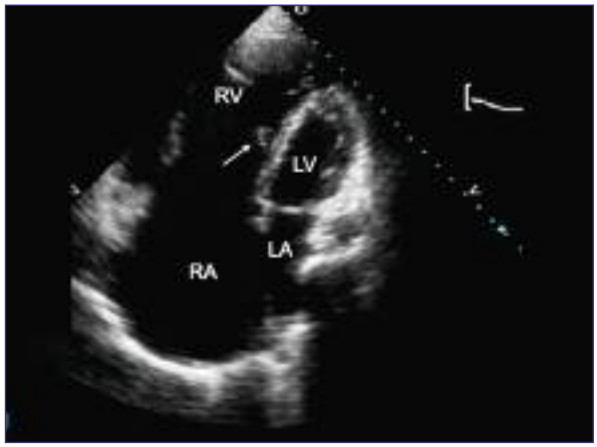

Figure 2. An apical four chamber view on transthoracic ECHO demonstrating apical displacement of the septal leaflet of the TV (arrow) with atrialization of the RV consistent with Ebstein's anomaly. RA, RV, left atrium [LA], left ventricle [LV].

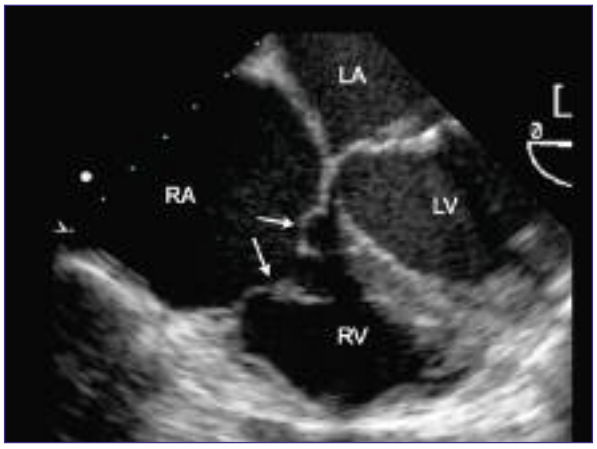

Figure 3. Midesophageal four chamber view on transesophageal ECHO demonstrating rheumatic involvement of the TV leaflets (arrows).

Rheumatic tv disease

A 32-year-old female correctional officer complained of SOB for the past two months. The cardiorespiratory examination was remarkable for a grade IV/VI pansystolic murmur at the LLSB of the heart, a pandiastolic murmur at the apex and a grade II/VI decrescendo diastolic murmur in the LLSB. The patient was referred for transesophageal ECHO, which revealed rheumatic tricuspid stenosis and regurgitation, mixed mitral valve disease and rheumatic aortic regurgitation (Figure 3). Complications from rheumatic heart disease arise two to three decades after an initial autoimmune response to a group A *Streptococcus* infection. Ten to twenty per cent of patients with rheumatic mitral and/or aortic valve disease concurrently have complications from rheumatic TV disease including tricuspid stenosis and regurgitation.[1] Although it is of foremost concern in developing countries, rheumatic TV disease should still be considered in the differential diagnosis of right-sided heart failure in North American rheumatic patients.[6]

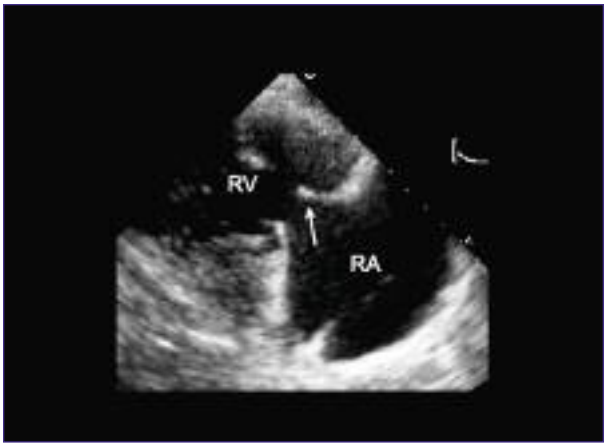

Figure 4. A RV inflow view on transthoracic ECHO demonstrating fixed, immobile leaflets of the TV (arrow), consistent with carcinoid heart disease.

Carcinoid heart disease

A 40-year-old man was diagnosed with carcinoid syndrome. Subjectively, he presented with a history of SOB for the past four months, diarrhea and abdominal pains. Physical examination was notable for an elevated JVP and hepatomegaly. ECHO revealed thickening and immobility of the TV leaflets with severe tricuspid regurgitation (Figure 4). Carcinoid heart disease is prevalent in > 50% of patients with carcinoid syndrome.[7] It is believed to result from factors secreted into the hepatic vein produced by a metastasis in the liver, initially originating from a GI tumour.[7] The serotonin metabolite, 5-Hydroxyindoleacetic acid (5-HIAA), may be the causal agent in the symptomatic fibrous plaque-like deposits found in cases of carcinoid heart disease. As a result, the right side of the heart is characteristically affected by thickening and shortening of the TV, as well as enlargement of the right atrium (RA) and RV. Treatment includes management of the primary tumour and replacement of the affected cardiac valves.[1,7]

TV papillary muscle rupture

A 70-year-old female patient was transferred to a tertiary care centre after failed thrombolytics for an inferior STEMI. Despite 15 L of supplemental oxygen, the patient remained hypoxic. ECHO confirmed a flail posterior TV leaflet, an inferior left ventricular (LV) wall motion abnormality, moderate RV systolic dysfunction and right to left shunting across a small atrial septal defect secundum (Figure 5). Due to the increased right atrial pressure from severe tricuspid regurgitation, there was right to left shunting resulting in hypoxia. Ten per cent of cases involving an inferior STEMI, from a right coronary artery occlusion, have RV involvement and may also result in ruptured tricuspid papillary muscles. Treatment generally consists of immediate surgical intervention to replace the TV since right-sided heart failure may follow.[8] Although rare, physicians should consider rupture of the TV papillary muscles in patients presenting with an inferior STEMI.

Carcinoid heart disease is prevalent in > 50% of patients with carcinoid syndrome.

PPH

A 37-year-old female presented with SOB for the past two months. On cardiorespiratory examination, the JVP was elevated at 5 cm above the sternal angle, with a grade II/VI pansystolic murmur detected in the lower left sternal border, and a dominant A wave. The S1 was normal with a loud P2 component. ECHO confirmed right ventricular dysfunction, flattening of the septum in systole and diastole consistent with RV pressure overload and a dilated pulmonary artery,
consistent with PPH (Figure 6).

PPH is characterized by an elevated pulmonary artery pressure > 25 mmHg at rest or 30 mmHg with exertion, in the absence of secondary causes.[9] Although the direct cause is unknown, studies suggest

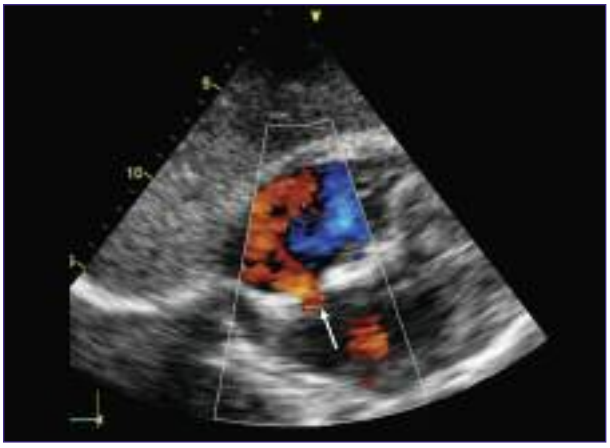

Figure 5. A subxiphoid view on transthoracic ECHO demonstrating left to right shunting across the interatrial septum on colour Doppler.

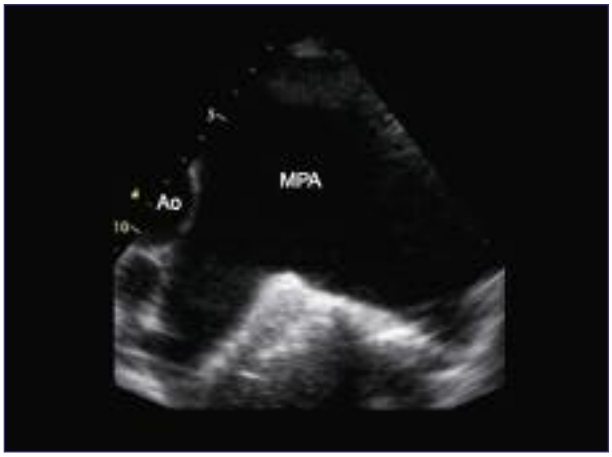

Figure 6. A short axis view at the level of the aortic valve, demonstrating an enlarged pulmonary artery consistent with pulmonary hypertension. Aorta [Ao], main pulmonary artery [MPA].

that appetite suppressants, inheritance, ingestion of cocaine or amphetamines and HIV infection may be contributing risk factors.[9-11] ECHO, CT and right heart catheterization are routinely performed in the diagnostic workup of patients with PPH. Treatment includes therapy to promote vasodilation, inhibition of platelet aggregation and/or anticoagulation and in severe cases lung transplantation.[9]

Conclusion

Although considered as the "forgotten valve," the TV should be carefully evaluated in patients with right-sided heart disease.

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References

1. Shah PM, Raney AA: Tricuspid Valve Disease. Curr Probl Cardiol 2008; 33(2):47-84.
2. Jassal DS, Picard MH: Infective Endocarditis: Management in the Era of Intravascular device. Chapter 13: Echocardiography. New York: Informa Healthcare USA, 2007.
3. Jassal DS, Weyman AE: Infective Endocarditis in the Era of Intracardiac Devices: An Echocardiographic Perspective. Rev Cardiovasc Med 2006; 7(3):119-29.
4. Dearani JA, Danielson GK: Congenital Heart Surgery Nomenclature and Database Project: Ebstein's Anomaly and Tricuspid Valve Disease. Ann Thorac Surg 2000; 69(4 Suppl):S106-17.
5. Bonow RO, Carabello BA, Chatterjee K, et al: ACC/AHA 2006 Guidelines for the Management of Patients With Valvular Heart Disease: A Report of The American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the 1998 Guidelines for the Management of Patients with Valvular Heart Disease) Developed in Collaboration With The Society of Cardiovascular Anesthesiologists Endorsed By The Society for Cardiovascular Angiography and Interventions and The Society of Thoracic Surgeons. J Am Coll Cardiol 2006; 48(3):1-148.
6. Carapetis JR, McDonald M, Wilson NJ: Acute Rheumatic Fever. Lancet 2005; 366(9480):155-68.
7. Kulke MH, Mayer RJ: Carcinoid Tumors. N Engl J Med 1999; 340(11):858-68.
8. Fujiwara K, Hisaoka T, Komai H, et al: Successful Repair of Traumatic Tricuspid Valve Regurgitation. Jpn J Thorac Cardiovasc Surg 2005; 53(5):259-62.
9. Runo JR, Loyd JE: Primary Pulmonary Hypertension. Lancet 2003; 361(9368):1533-44.
10. Barst RJ, McGoon M, Torbicki A, et al: Diagnosis and Differential Assessment of Pulmonary Arterial Hypertension. J Am Coll Cardiol 2004; 43(12 Suppl S):40S–47S.
11. Jassal DS, Sharma S, Maycher B: Pulmonary Hypertension. Emedicine 2004. Http://Emedicine.com.