

# Orthotopic Cardiac Transplantation for Chagas Cardiomyopathy in Australia

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## What is Chagas Disease?

- Chronic, systemic parasitic condition caused by the protozoa *Trypanosoma cruzi* (*T. cruzi*).
- Endemic in many Latin American countries (Figure 1).
- Acute infection can either be asymptomatic, or present with fever and lymphadenopathy.
- Chronic infection primarily manifests as dilated cardiomyopathy (DCM) and digestive tract disease (megaesophagus or megacolon).
- Can also lead to arrhythmias and sudden cardiac death (SCD).
- Transmitted to humans and animals by a large, blood sucking Triatominae bug via excretion of the parasite in infected insect faeces (Figure 2).
- Diagnosis is made using serologic tests specific for *T. cruzi* IgG antibodies using at least two different methods ie. Enzyme-linked immunosorbent assay (ELISA) or indirect immunofluorescence (IF).
- PCR can be used to detect acute or congenital Chagas cases. Chronic infection will have minimal active parasitemia.

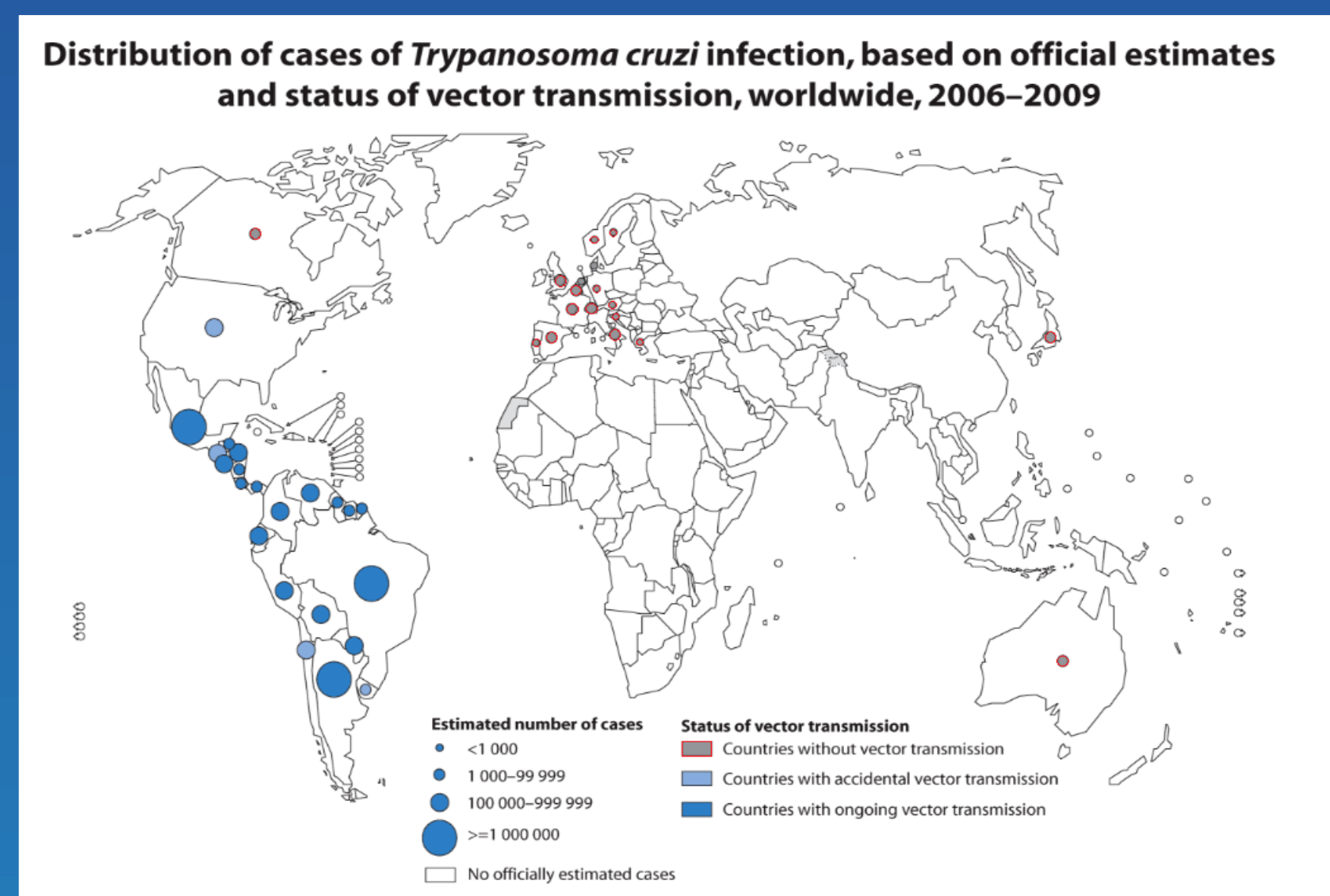


Figure 1: Geographic Distribution of *T. cruzi* infection<sup>1</sup>

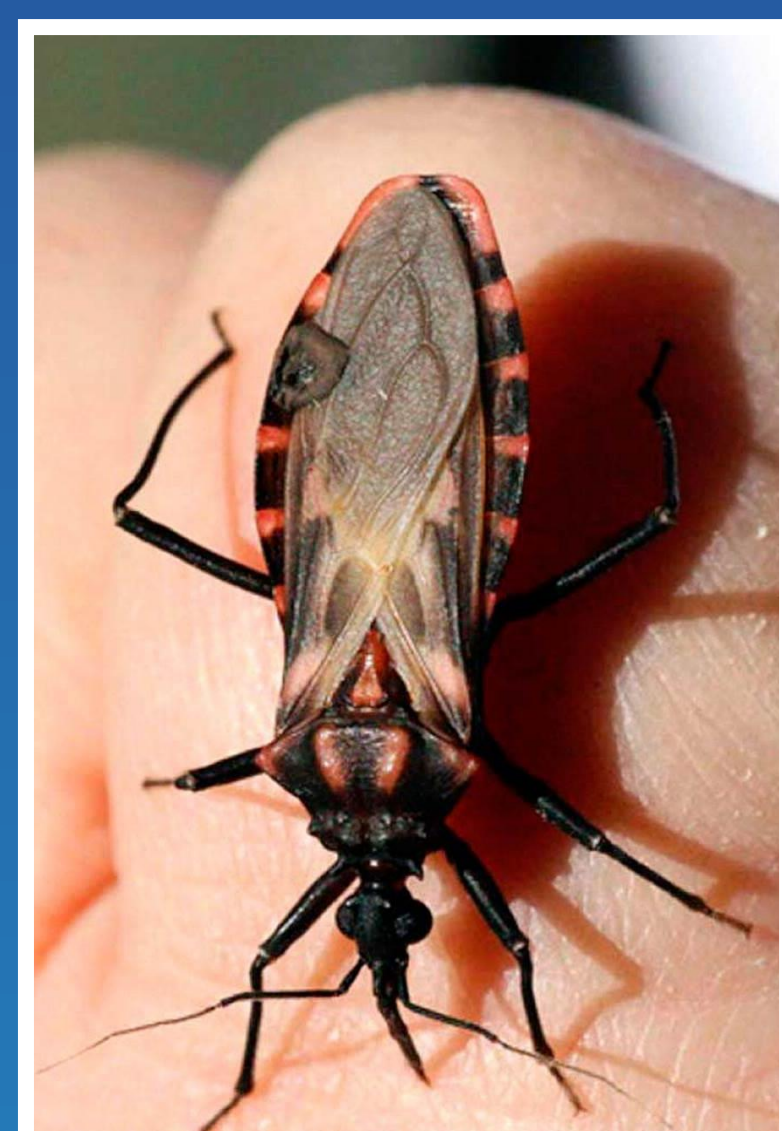


Figure 2: Vector: Triatominae bug, a subfamily of Reduviidae.<sup>6</sup>

## Case Presentation

A 54-year-old male of Central-American descent with a diagnosis of idiopathic dilated-cardiomyopathy was referred to a tertiary referral center for cardiac transplantation assessment. He had no other medical co-morbidities and had been living in Australia for 26 years. At initial presentation three years prior, the left ventricular ejection fraction was 15%. Conventional cardiomyopathy screening was negative. Cardiac MRI showed partial to full thickness scarring of both the lateral and apical left ventricular walls. Coronary angiography revealed minor coronary artery disease only.

## Investigation and Management

During further assessment, *T. cruzi* IgG-EIA serology was found to be positive, suggesting previous exposure. The patient was initially managed with conventional heart failure therapies but he subsequently presented with an out of hospital cardiac arrest and a secondary prevention ICD was inserted. He made good recovery from the cardiac arrest and remained stable for two years on medical therapy until severe heart failure led to his listing for cardiac transplantation. Transthoracic echocardiography at this time revealed a severe, dilated cardiomyopathy with an ejection fraction of 20%, severe mitral regurgitation and mild right ventricular dysfunction.

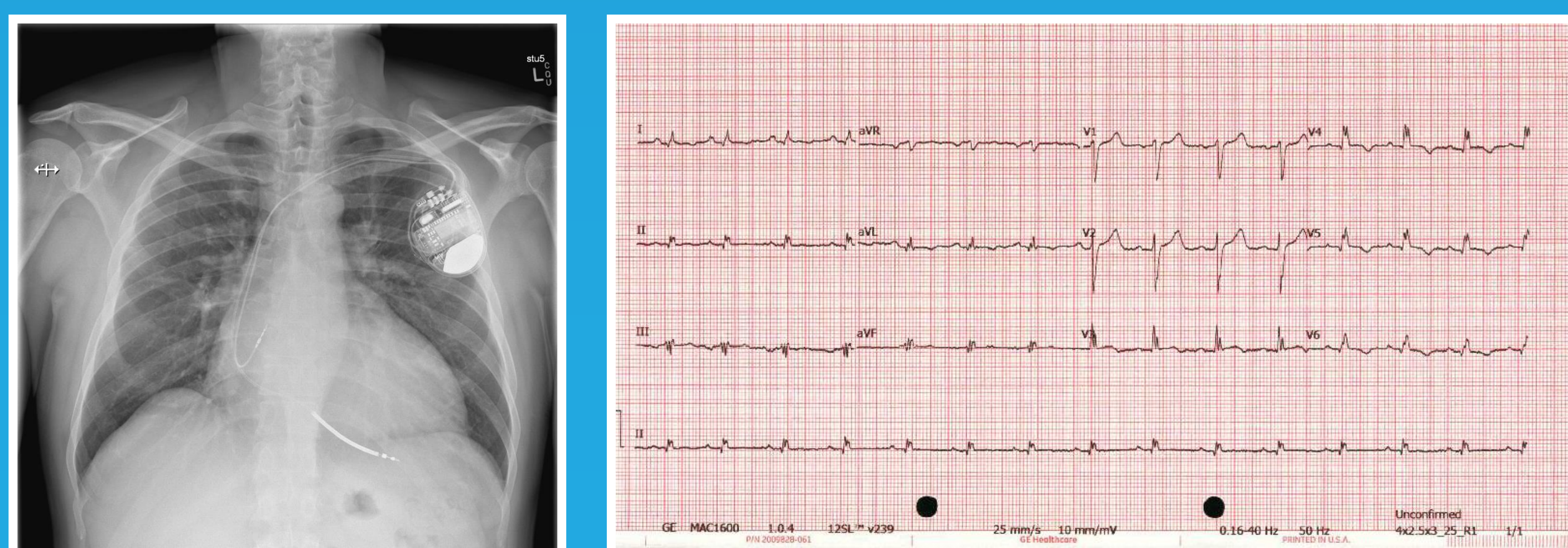


Figure 3: Chest x-ray showing cardiomegaly and dual chamber device in-situ, and pre-transplant electrocardiogram

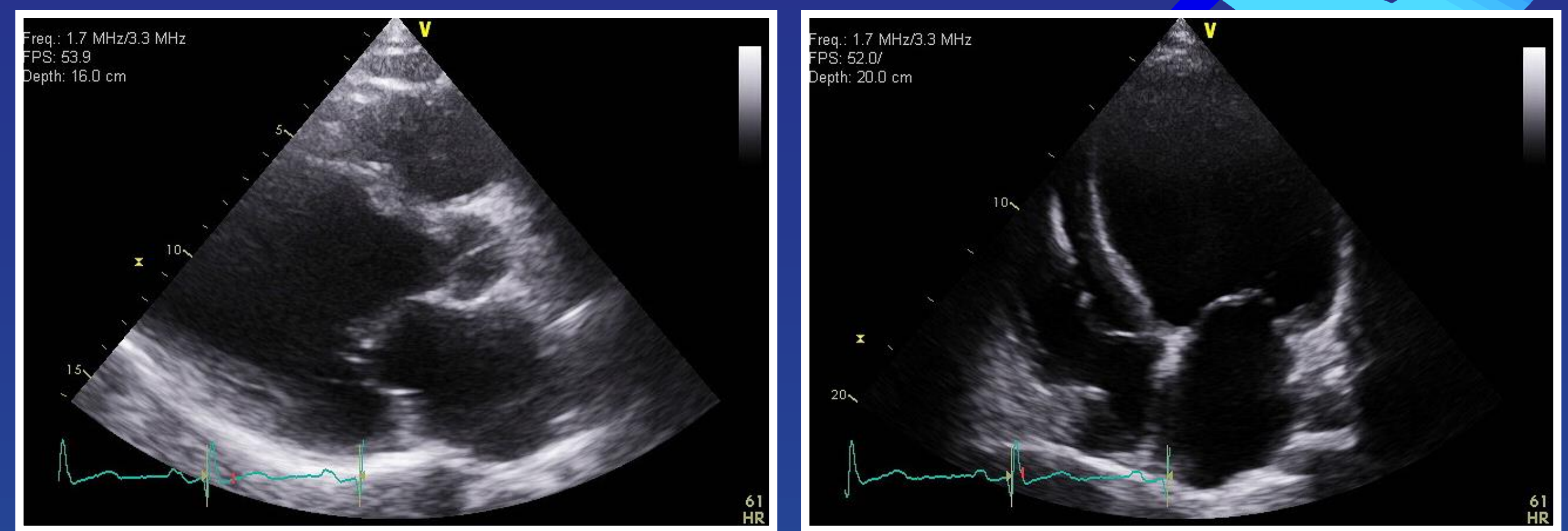


Figure 3: TTE (a) Parasternal long axis view showing a dilated left ventricle (left image); (b) Apical-4-chamber view showing severely dilated left ventricle and right ventricular lead insitu (right image)

The patient underwent successful cardiac transplantation. Histology of the explanted heart revealed diffuse patchy myocarditis involving both left and right ventricles. There was a lymphocyte predominant infiltrate, and significant numbers of eosinophils, a small number of plasma cells but no giant cells or granuloma formation. No parasites were visualised which is consistent with chronic Chagas cardiomyopathy.

## Progress

Post transplantation, weekly blood screening for Chagas reactivation was initiated, with specimens sent to CDC Atlanta USA for *T. cruzi* PCR. At one month post transplantation, *T. cruzi*-PCR testing became positive and he received sixty days therapy of Benznidazole 150mg twice-daily obtained from the WHO, Geneva. *T. cruzi*-PCR became negative in time, and all cardiac biopsies have been negative for Chagas recurrence. He had no adverse reactions to Benznidazole whilst on therapy. One year post treatment he remains relapse-free.

## Discussion

- Chagas cardiomyopathy is a rare cause of heart failure in Australia.
- Consider Chagas disease in patients presenting with heart failure from endemic regions (it is estimated that we have ~1928 infected residents in Australia).<sup>5</sup>
- Acute infection is predominately a self limiting febrile illness, thus the majority patients infected are unaware.<sup>2</sup>
- 30-40% of infected individuals will develop chronic Chagas disease, with onset of disease approximately 10-30 years after initial infection.<sup>2</sup>
- DCM is a late manifestation and is associated with left ventricular apical aneurysms, ventricular ectopy & ventricular tachycardia.<sup>3</sup>
- Diagnosis of Chagas disease is made by detection of parasitemia in acute infection, or serologic tests for *T. cruzi* in chronic infection.<sup>2</sup>
- Cardiomyopathy treatment includes ACE inhibitors and amiodarone or device therapy for arrhythmias. Cautionary use of beta blockers is recommended given the risk of bradyarrhythmias.<sup>2</sup>
- The only effective *T. cruzi* therapies are benznidazole or nifurtimox.<sup>2</sup>
- Multi-disciplinary involvement at an early stage (pre-transplant) is suggested for Chagas cardiomyopathy.
- Cardiac transplantation is indicated for end-stage cardiomyopathy due to Chagas, with reasonable post-transplant outcomes.<sup>4</sup>
- Immunosuppressed patients are at risk of Chagas reactivation and frequent screening for this is crucial.<sup>2</sup>

## References

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