I'M IN TWO HEARTS ABOUT THIS DIAGNOSIS!

Case presentation
61 year old male patient with a history of heterotropic heart transplant (HHT) presented to ED by ambulance following two syncopal episodes. Ambulance crew reported abnormal ECG findings.
The patient had felt hot, sweaty and clammy with palpitations preceeding both syncopal events. He had chest and epigastric pain in the past 24 hours. Syncopal events witnessed by partner who reported a grey and clammy appearance of the patient.
During the A&E review, the patient reported ongoing chest discomfort with no associated symptoms.

Examination findings:
Pulse: 90bpm
Blood pressure: 120/94
SpO2: 98% RA
RR: 18/min
Apex beat palpated on the right precordium
Chest: Bilaterally clear, equal air entry.

Investigations
A basic blood panel, XR chest and ECG

Point-of-care echocardiogram
This was crucial in the diagnosis of this patient. Viewing the two hearts in real time allowed the clinicians to **visualize a fibrillating native heart and the donor heart being in sinus rhythm**. Severe systolic dysfunction was noted of the native heart. Along with this, the formal Echo showed severe AR and severe MR

Diagnosis
Syncope secondary to Electrical Storm generated from native heart. VT propagation to donor heart.

Initial Management
The patient was promptly transferred to resuscitation area. Given the high risk of sustained ventricular tachycardia/fibrillation, amiodarone infusion was initiated as an antiarrhythmic, and preparations for potential cardioversion were made. The on-call cardiology team was consulted for urgent evaluation, with plans for direct current cardioversion (DCCV) if needed. The patient was closely monitored, and subsequently transferred to intensive care.

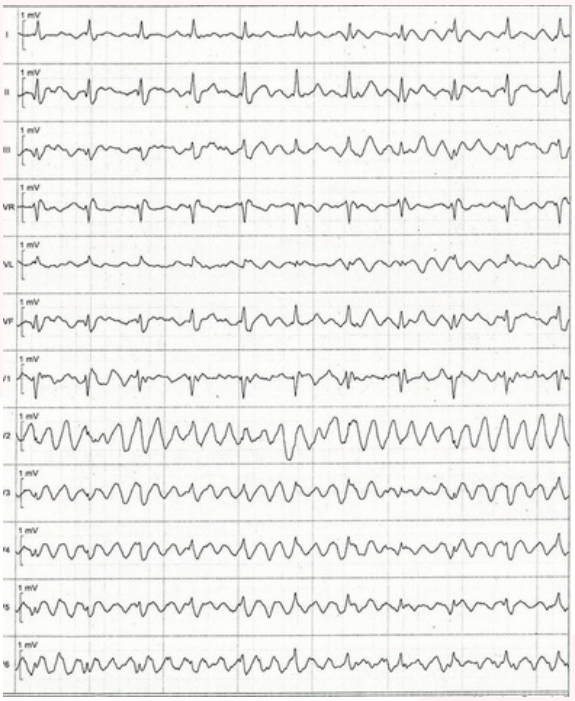

Fig. 1: Limb leads: Sinus rhythm from the transplanted heart was seen in the right sided leads
Precordial leads: Ventricular fibrillation was being picked up from the native heart on the left lateral leads. There is however an overlap of these rhythms seen on the ECG here.

Diagnostic Challenges
> Clinicians need to think outside the box. Syncope secondary to conventional diagnoses did not apply to this patient due to the rarity of this condtion
> A clinical mismatch with the underlying pathology was unexpected. This can lead to delays in care or missed recognition
>ECG misinterpretation can lead to incorrect diagnosis as the precordial leads could have been read as arterfact. Bedside POCUS Echo led to additional information showing a tachycardic native heart whilst donor heart being in sinus rhythm.
>Ventricular tachycardia from the native heart was propagating to the donor heart. This was the likely cause of the patient's symptoms (syncope)

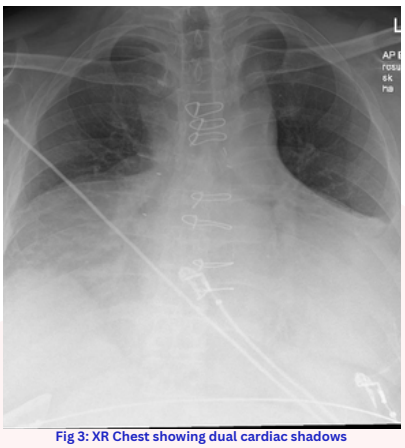

Fig 3: XR Chest showing dual cardiac shadows

VENTRICULAR TACHYCARDIA

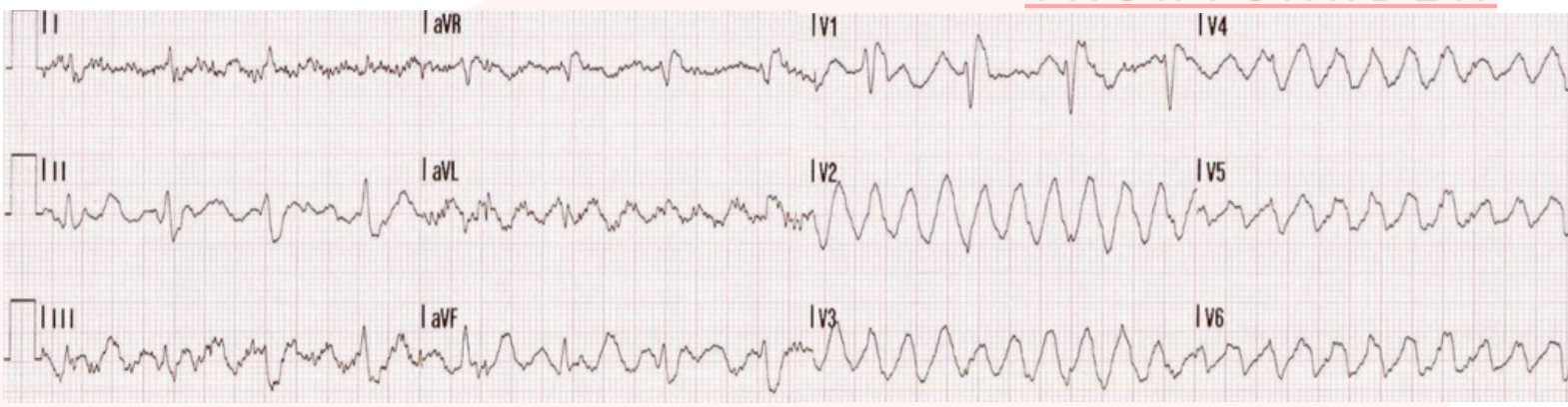

Fig 2: Ambulance ECG showing ventricular tachycardia in the left lateral leads mixed with sinus rhythm in the right sided limb leads. This extends to V1 as well.

ITU Management for Rhythm control
- The patient was transferred to transplant centre ITU, where the amiodarone infusion was continued with addition of IV magnesium. Despite this the patient's native heart remained in VT.
- This prompted the need for direct current cardioversion for which transoesophageal echocardiography (TOE)-guided DCCV was chosen, allowing real-time imaging to ensure precise electrode placement and minimise complications.
- The first attempt at DCCV did not achieve lasting success, as the patient reverted to VT shortly after. Consequently, a second round of DCCV was performed. Following this second shock, the patient remained in sinus rhythm for a brief period, but VT recurred.
- **Resolution:** Given the persistence of VT, lidocaine infusion was started and another round of TOE guided DCCV helped convert rhythm to sinus.
- **Discharge:** Amiodarone infusion was stopped and after a discussion in Electophysiology Multidisciplinary meeting along with replacing patients regular bisoprolol with oral sotalol and addition of oral mexilitine. Patient was stepped down to a cardiac ward where lidocaine infusion was stopped. After a brief observation period, the patient was discharged with close follow-up. A lower respiratory tract infection was thought to have instigated the intial arrhythmia.

Literature Review
- Heterotropic Heart Tranplants, once a common procedure in the 1980s and early 1990s for patients with high pulmonary vascular resistance or donor-recipient size mismatch, is now exceedingly rare with very few active cases remaining worldwide. (1)
- In HHT, both the donor and native hearts function in parallel, this can present diagnostic challenges, particularly in the context of arrhythmias. Ventricular tachycardia (VT) originating from the native heart is a recognized but uncommon phenomenon, and in most cases does not lead to systemic compromise due to continued output from the donor heart. (2)
- Sometimes, the arrhythmia in the native heart can lead to syncope or ischaemic symptoms. (3)
- Electrocardiogram interpretation is often confounded by electrical activity of dual morphology, which requires advanced imaging and specialist evaluation to accurately localize the rhythm origin (4)
- Antiarrhythmic Drugs: Amiodarone is frequently used as a first-line agent for rhythm control in patients with sustained VT due to its ability to stabilize the cardiac rhythm in both donor and native hearts (5). However, as seen in this case, the long-term use of amiodarone can lead to adverse effects, and alternatives like sotalol or mexiletine may be used if the arrhythmia persists or the patient develops side effects (6)

Learning points for the Emergency Medicine Physician

- Understand HHT Physiology: Heterotopic heart transplant means two hearts in one chest — both native and donor hearts can generate electrical activity and influence haemodynamics.
- Beware of ECG Misinterpretation: VT in the native heart may resemble artefact or be misread as benign. Always correlate with history and bedside imaging.
- POCUS is a Game-Changer: Point-of-care echo revealed a native heart in VT and a donor heart in sinus rhythm — crucial for diagnosis.
- Mismatch Between Presentation and Pathology: The patient appeared stable, but was in sustained VT/VF in native heart. Don't rely solely on vitals or appearance.
- Recognise Imaging Clues: A double cardiac shadow on chest X-ray hinted at heterotopic transplant — an easy-to-miss but vital clue.
- Treat the Right Heart: Interventions should be directed at the heart causing the problem. Don't shock blindly — identify the arrhythmic source. TOE guided cardioversion is ideal.
- HHT is Rare — But It Happens: These cases are unusual, but not extinct. Be prepared for complex presentations that don't fit common patterns.

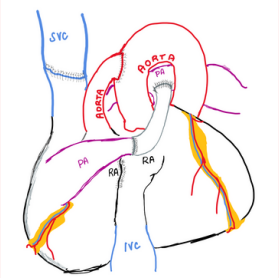

Fig 4: Schematic representation of heterotropic heart transplant

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