Isolated Noncompaction of Right Ventricular Myocardium and Arrhythmia

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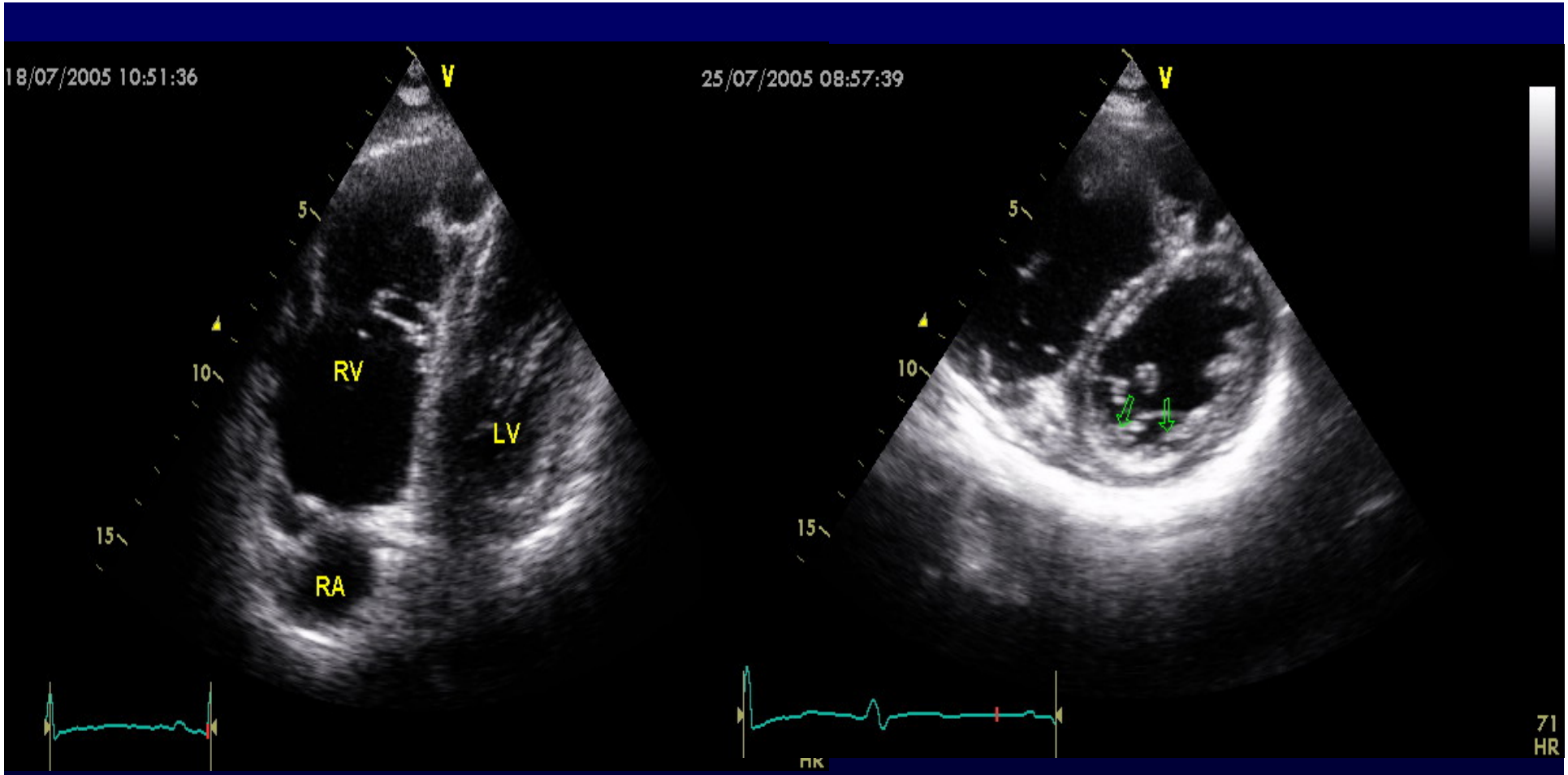
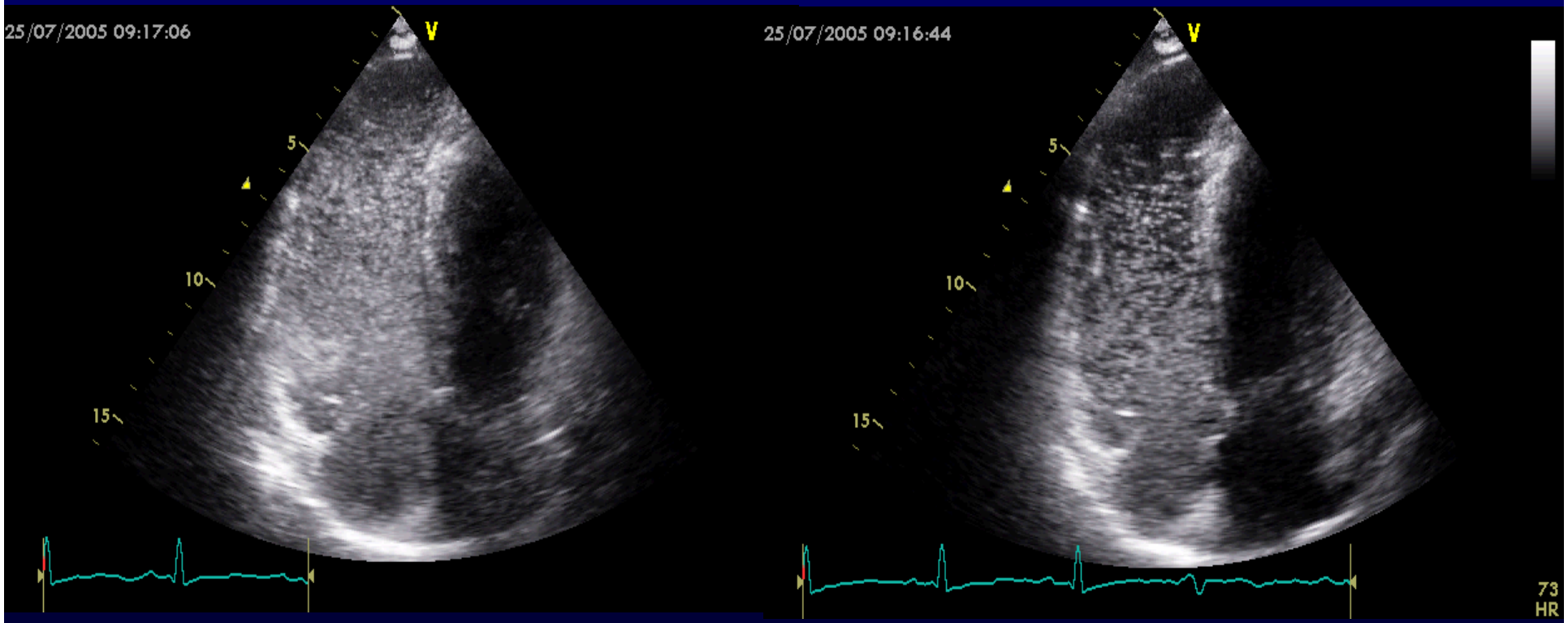
- A 28 years-old woman
- Palpitations, chest distress for 2 months
- History of syncope history with ventricular tachycardia
- No embolism event
- Propafenone: oral dose 150mg tid for 2 months
- No family history of cardiomyopathy

- **HR: 64 bpm, BP: 94/64mmHg**
- **Jugular venous pressure was normal**
- **No cardiac murmur**
- **A chest X ray showed marked cardiomegaly**
- **EKG: PVC, VT (Fig 3, 4)**
- **UCG: right ventricular enlargement 4.7 X 9.0 cm, INVM (Fig 1) , a local thin wall at the apex of right ventricle (0.2 cm)**
- **MRI: Fig 2**
- **Holter: Fig 5**



A: the 4-chamber view B: short-axis view

Fig 1 Two-dimensional echocardiograms showing the thick trabeculations and deep intertrabecular recesses in the right ventricular posterior wall and apex



- **Fig 2 H_2O_2 imaging showed H_2O_2 from the right ventricular cavity into the deep intertrabecular recesses during diastole**

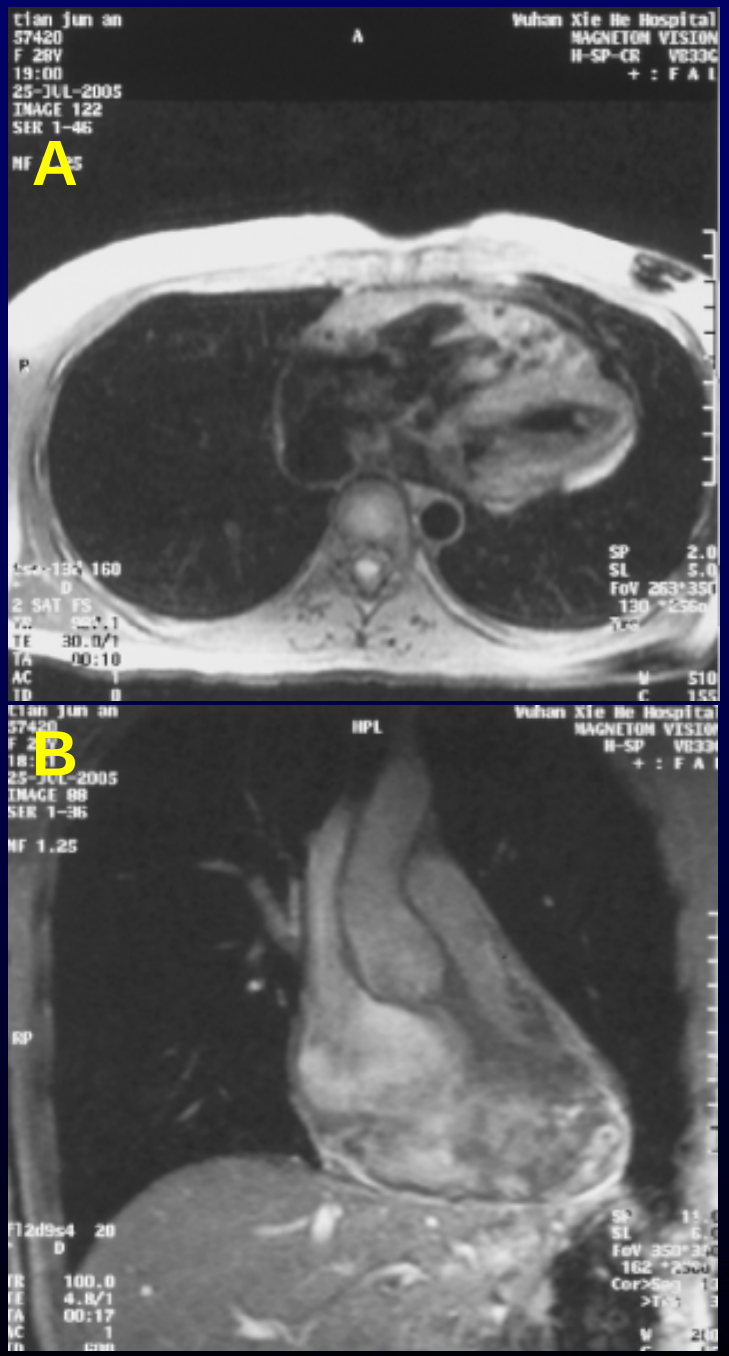


Fig 3

MRI showing a typical honeycombing appearance, excessively prominent trabeculations and deep intertrabecular recesses in the right ventricle

A: a transaxial plane

B: a coronal plane, from a steady state with free precession cine

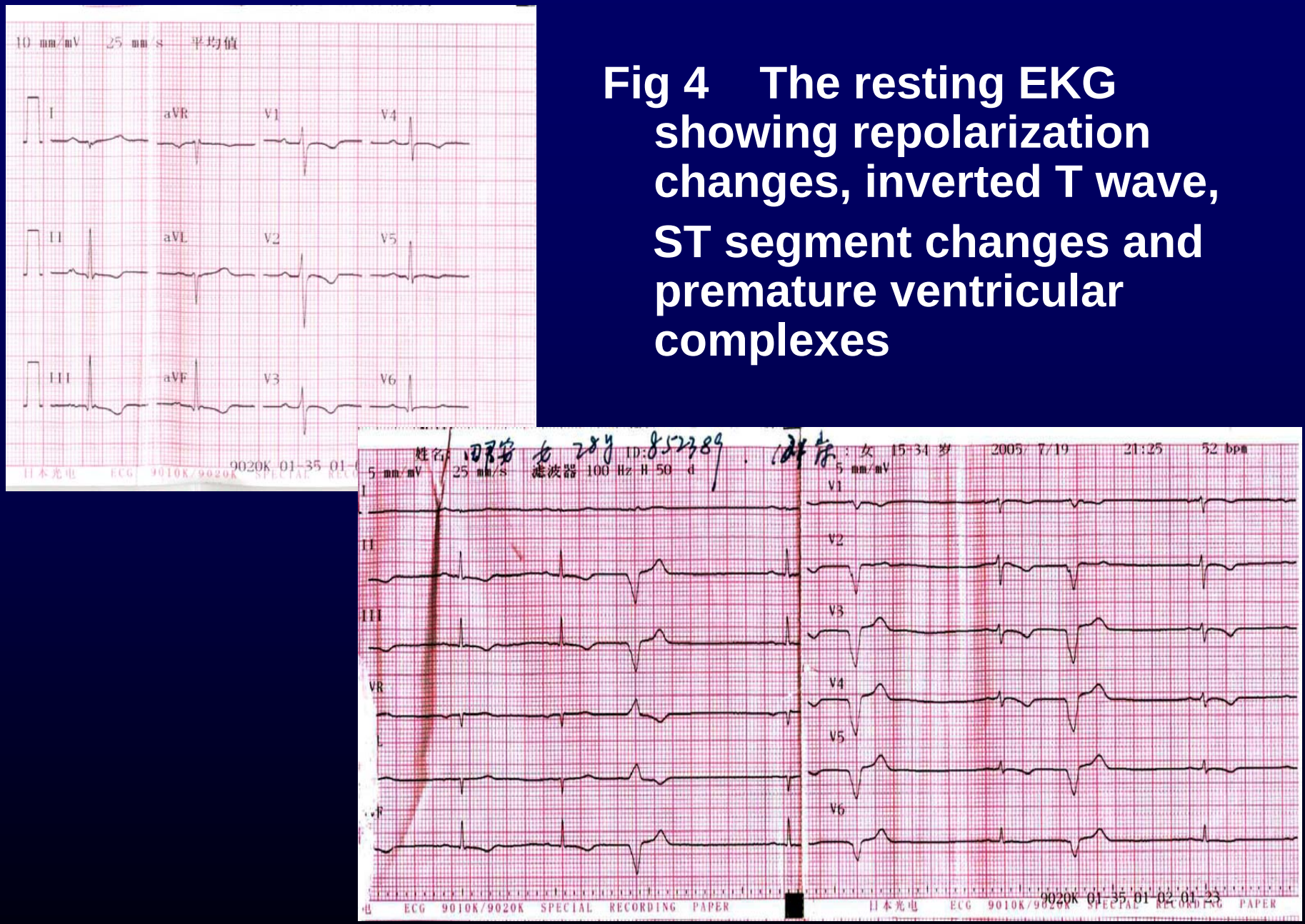


Fig 4 The resting EKG showing repolarization changes, inverted T wave, ST segment changes and premature ventricular complexes

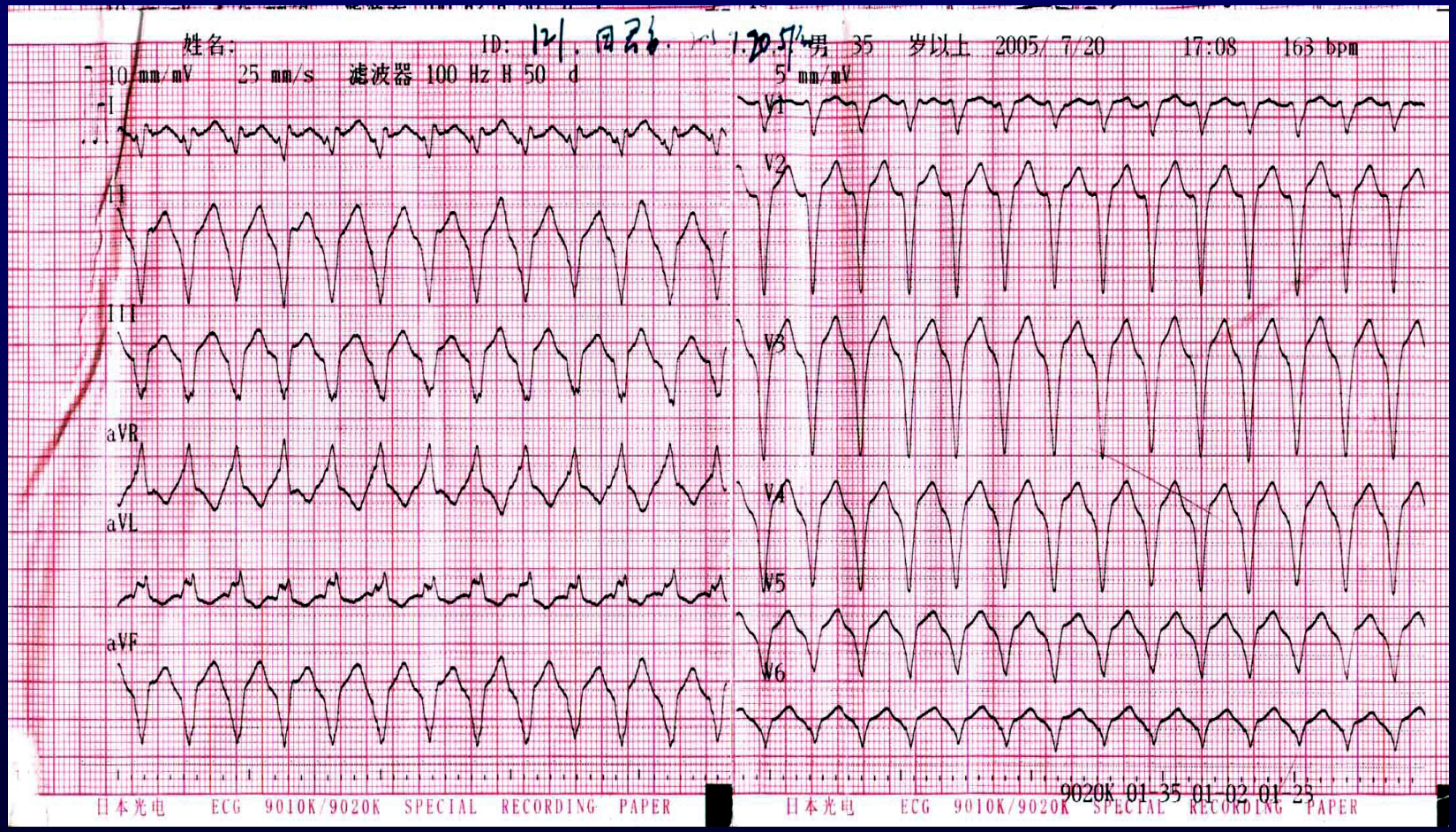
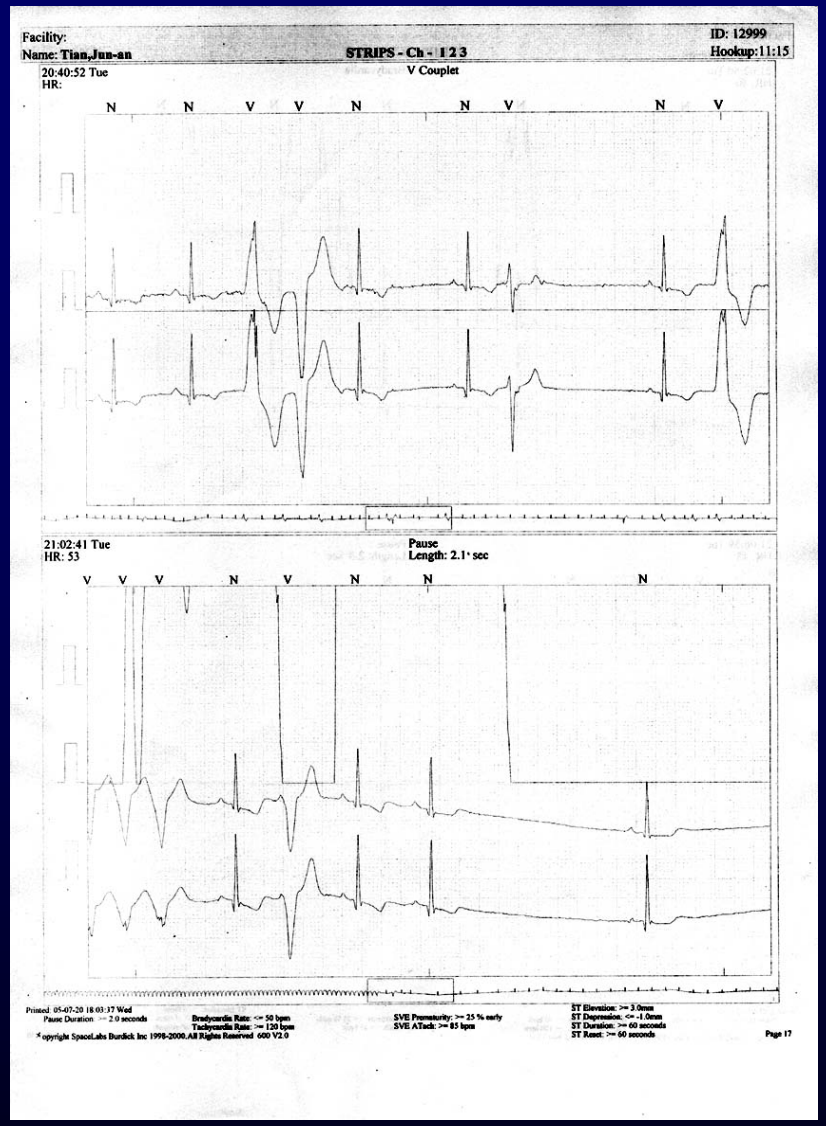


Fig 5 Symptoms (dizziness、amaurosis and a blurred vision) occurring during sustained ventricular tachycardia. VT stopped after IV administration Amiodarone



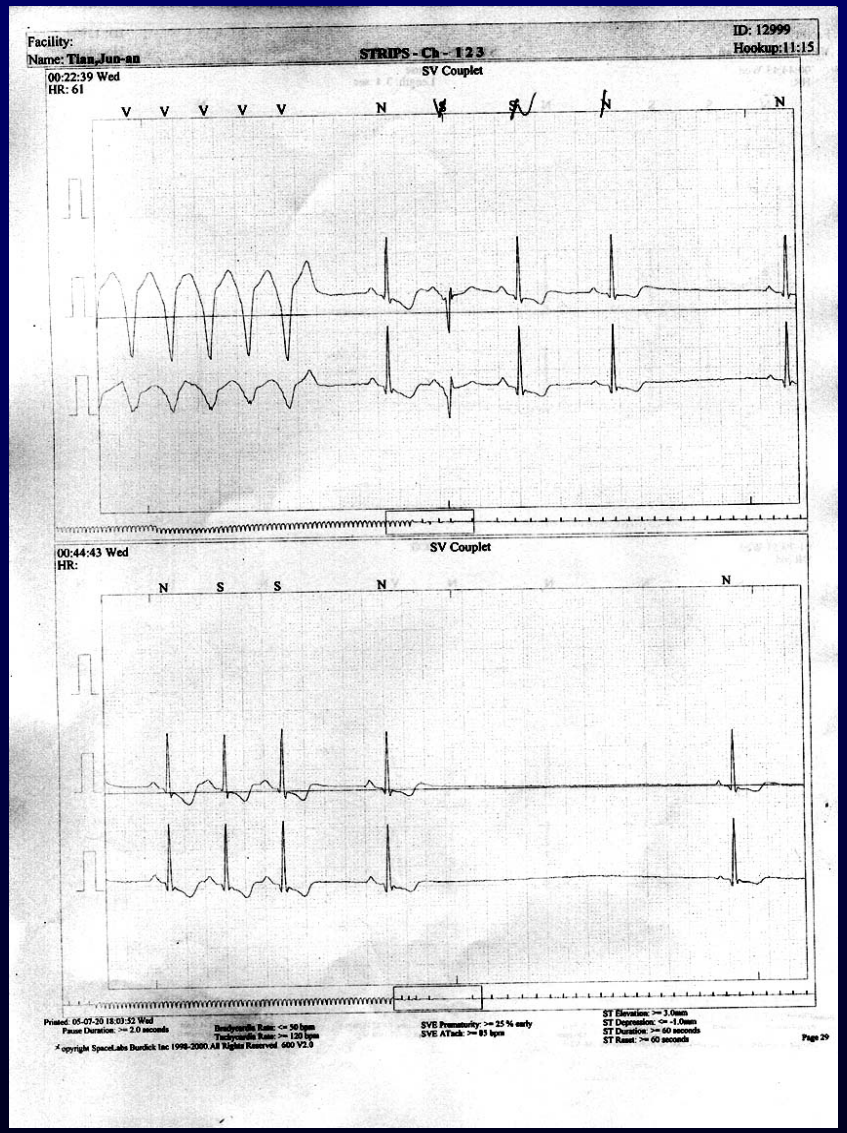


Fig 6 Holter：sinus rhythm, sinus arrest (3.6 s)

Sustained VT (lasted 52min), multifocal and multiform PVC, PAC et al.

Diagnosis

Primary genetic cardiomyopathy

- **Isolated noncompaction of right ventricle**
- **Life-threatening arrhythmia**
- **Sustained VT**
- **Transient sinus arrest (drug-related)**

Management

- Stopped all oral anti-arrhythmia bullet
- During hospitalization, she had a sustained VT stopped by IV administration of amiodarone
- Radiofrequency ablation of ventricular tachycardia (apex of the right ventricle)

Follow-up

- One year, no syncope
- Sometimes her EKG showed PVCs
- Aspirin 100mg qd

(Regular monitoring of INR is difficult for patient, so she is no on anticoagulants)

Discussion

- **Isolate Noncompaction of the Ventricular Myocardium (INVM) is a primary genetic cardiomyopathy thought to be caused by arrest of normal embryogenesis of the endocardium and myocardium**
- **The left ventricle is usually affected, but biventricular and right ventricular noncompaction have been reported**

- **Several authors dispute the existence of right ventricular noncompaction, because of difficulty in distinguishing normal variants in the highly trabeculated right ventricle from the pathological noncompaction.**

- **For this patient, it is a pathological change, including enlargement of the right ventricle, multiple prominent trabeculations with deep intertrabecular recesses and life-threatening ventricular arrhythmia**

- For diagnosis, we thought it is potentially useful for H_2O_2 imaging as a supplement to 2D UCG in the assessment of noncompaction
- It was challenge knowing how to treat this patient.

  For sinus arrest, we thought it might be related to the propafenone so we stopped the drug and observed the patient

  For VT, we chose the ablation therapy

Question

What is the optimal therapy for this case?