Isolated Noncompaction of Right Ventricular Myocardium and Arrhythmia

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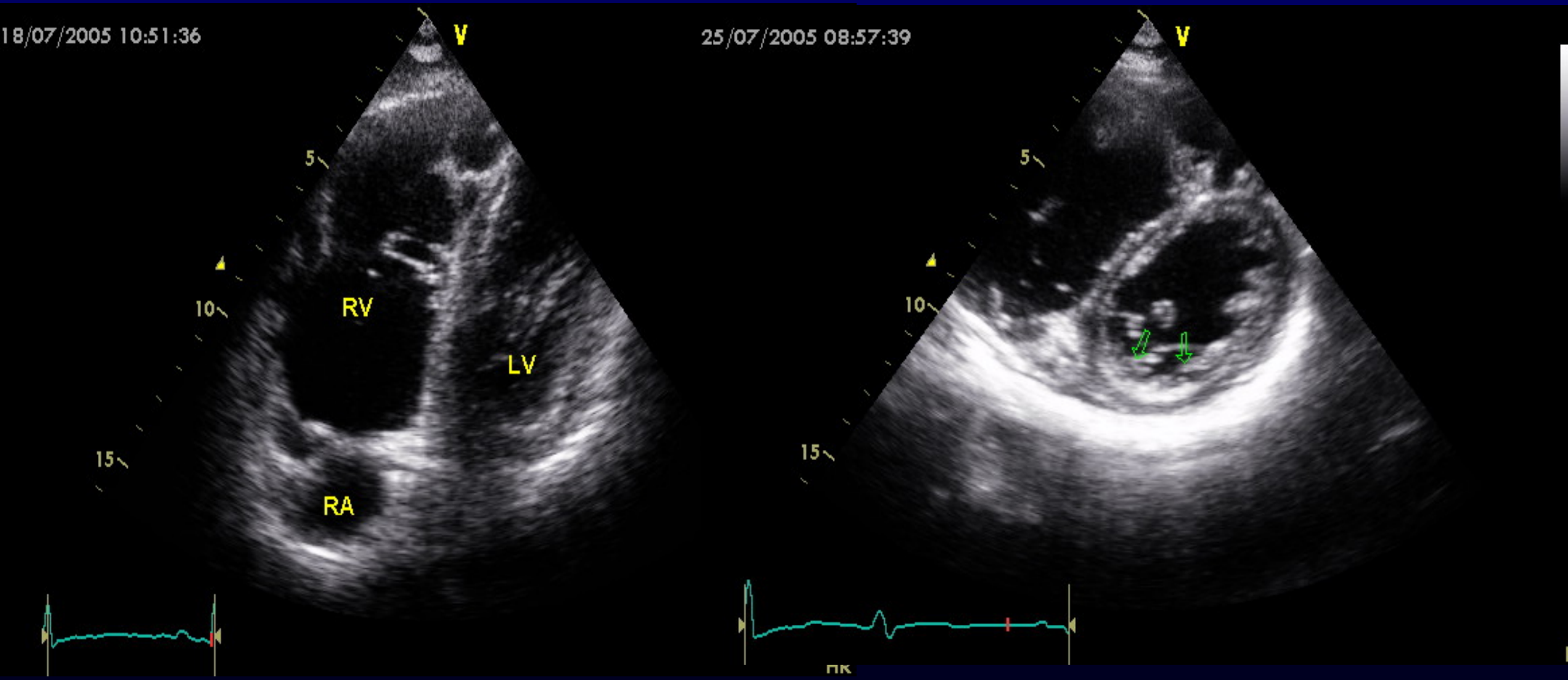
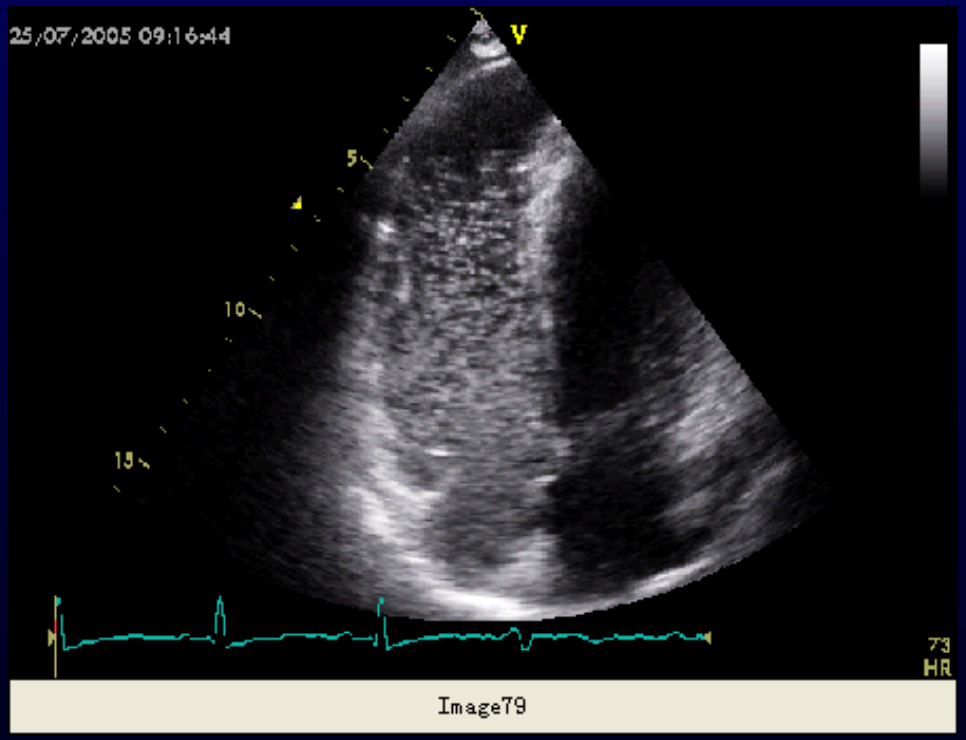
- A 28 years-old woman
- Palpitation, chest distress for 2 months
- Had a syncope history with ventricular tachycardia
- No embolism event
- Propafenone: oral dose 150mg tid for 2 months
- No family history of cardiomyopathy

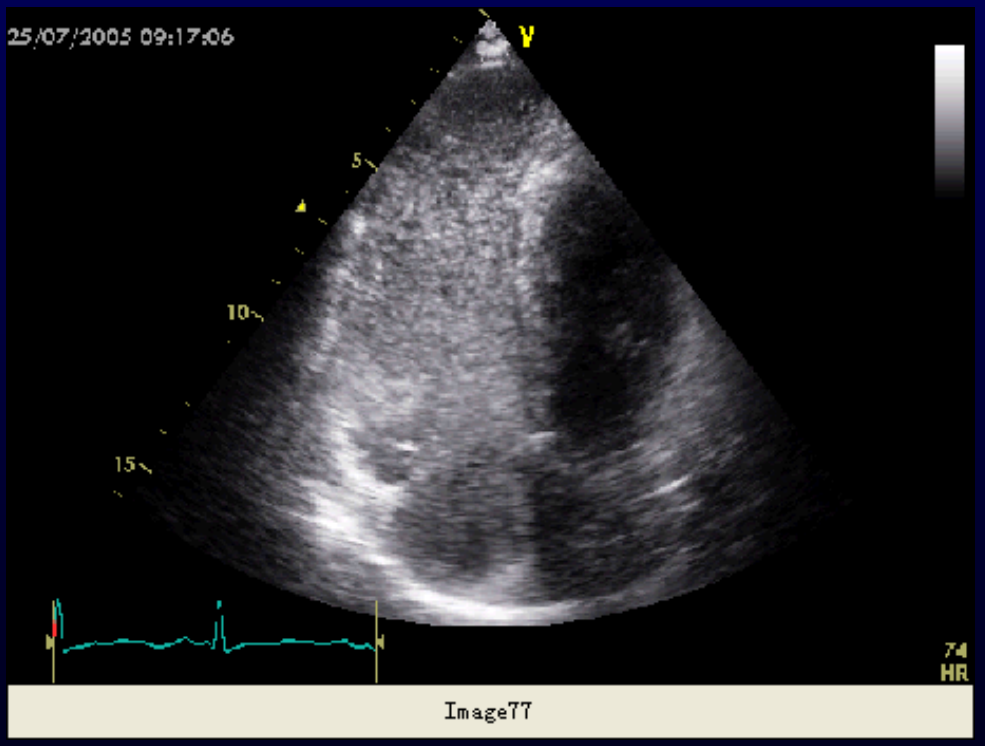
- HR: 64 b/m, BP: 94/64mmHg
- Jugular venous pressure was normal
- No cardiac murmur
- A chest X ray showed marked cardiomegaly
- EKG: nonspecificic ST-Tchanges, PVC, VT (Fig,3,4)
- UCG: right ventricular enlargement 4.7 X 9.0 cm, INVM (Fig 1) , a local thin wall at the apex of right ventricle (0.2 cm)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



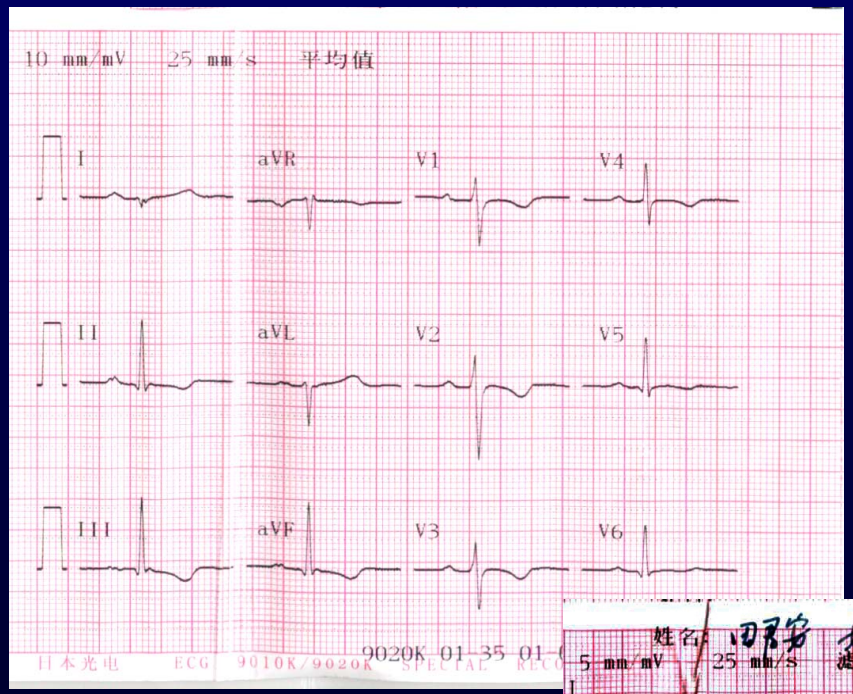


- Fig2 H_2O_2 imaging showed H_2O_2 from the right ventricular cavity into the deep intertrabecular recesses during diastolic

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



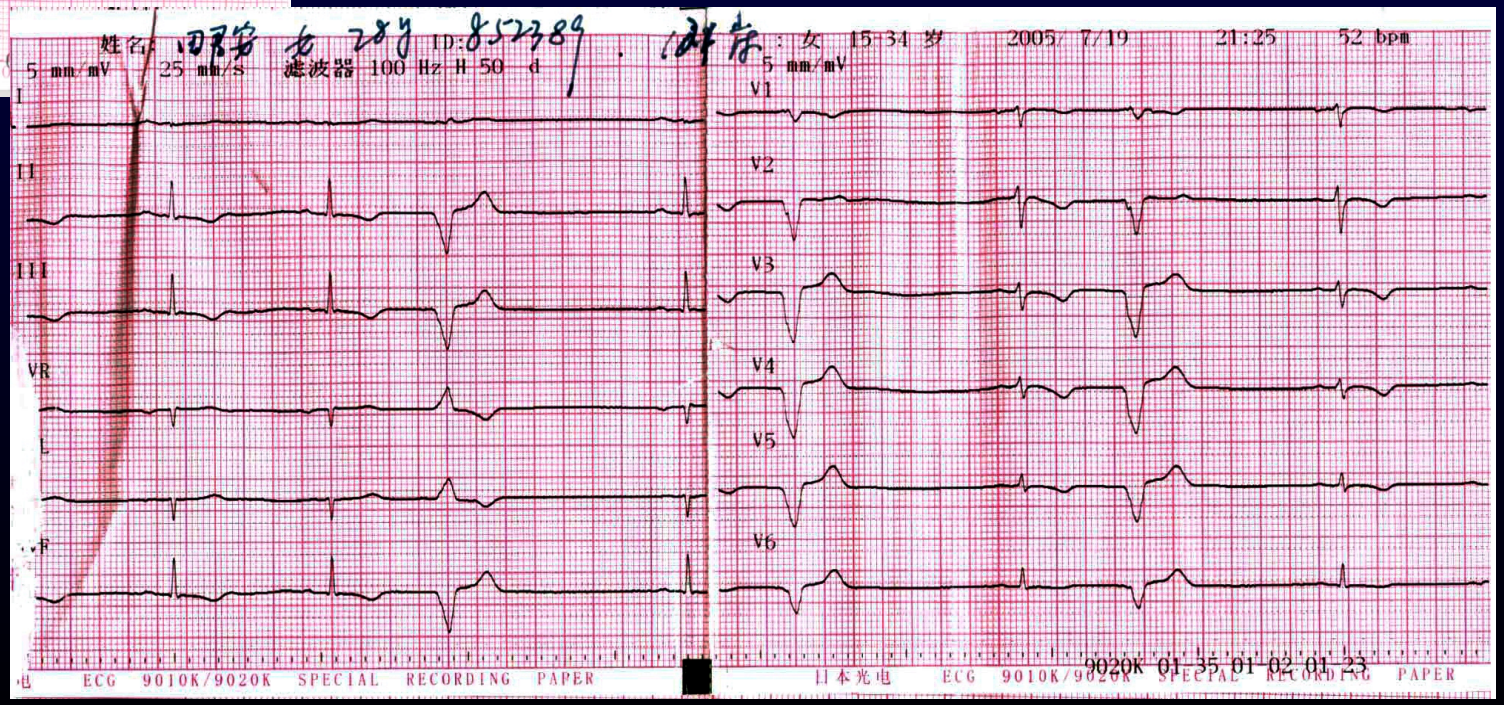


Fig 4 The resting EKG showing repolarization changes, inverted T wave, ST segment changes and premature ventricular complexes

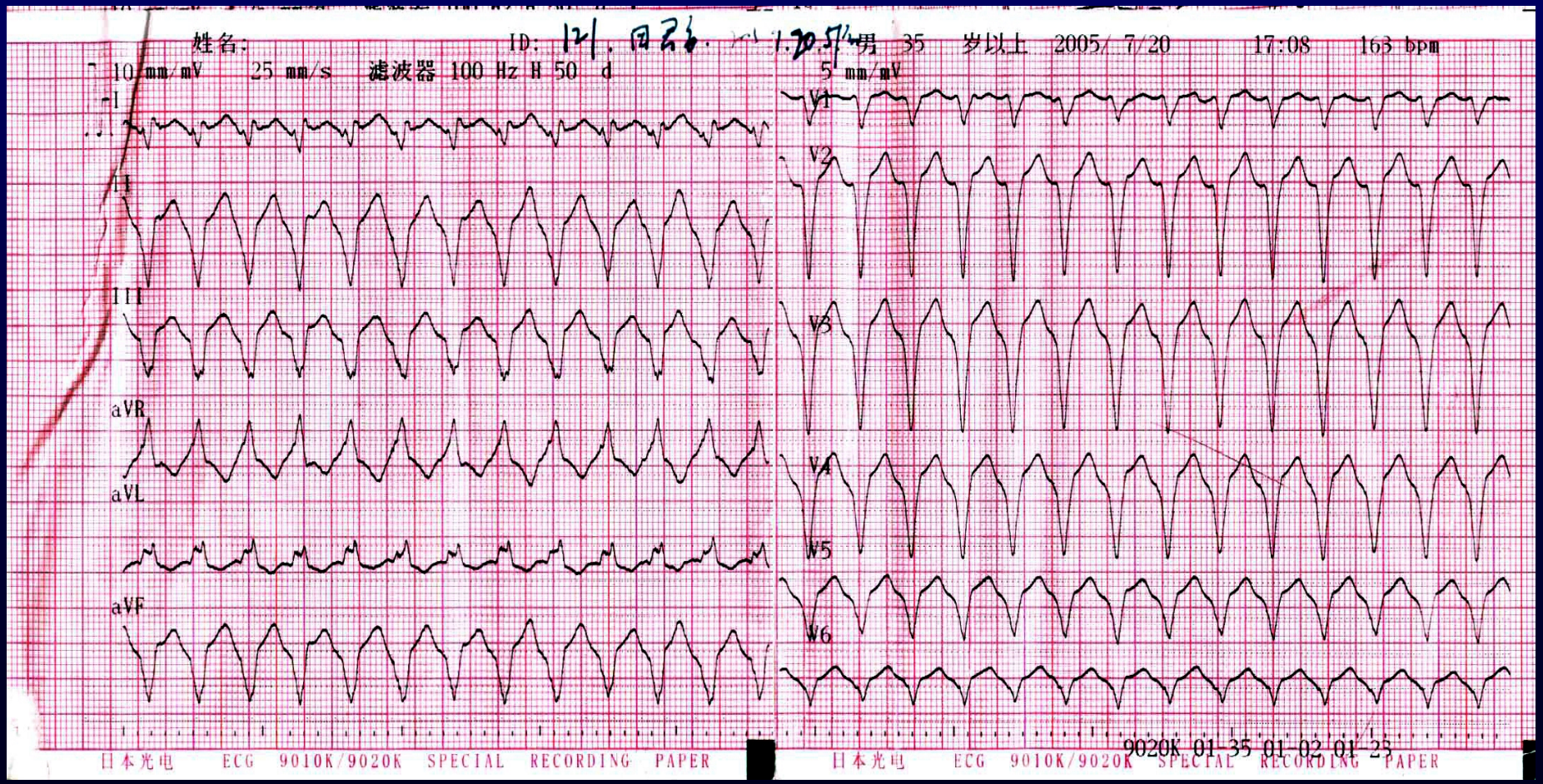


Fig 5 Symptoms (dizziness、amaurosis and a blurred vision) occurring during sustained ventricular tachycardia. Stop VT after IV administration Amiodarone

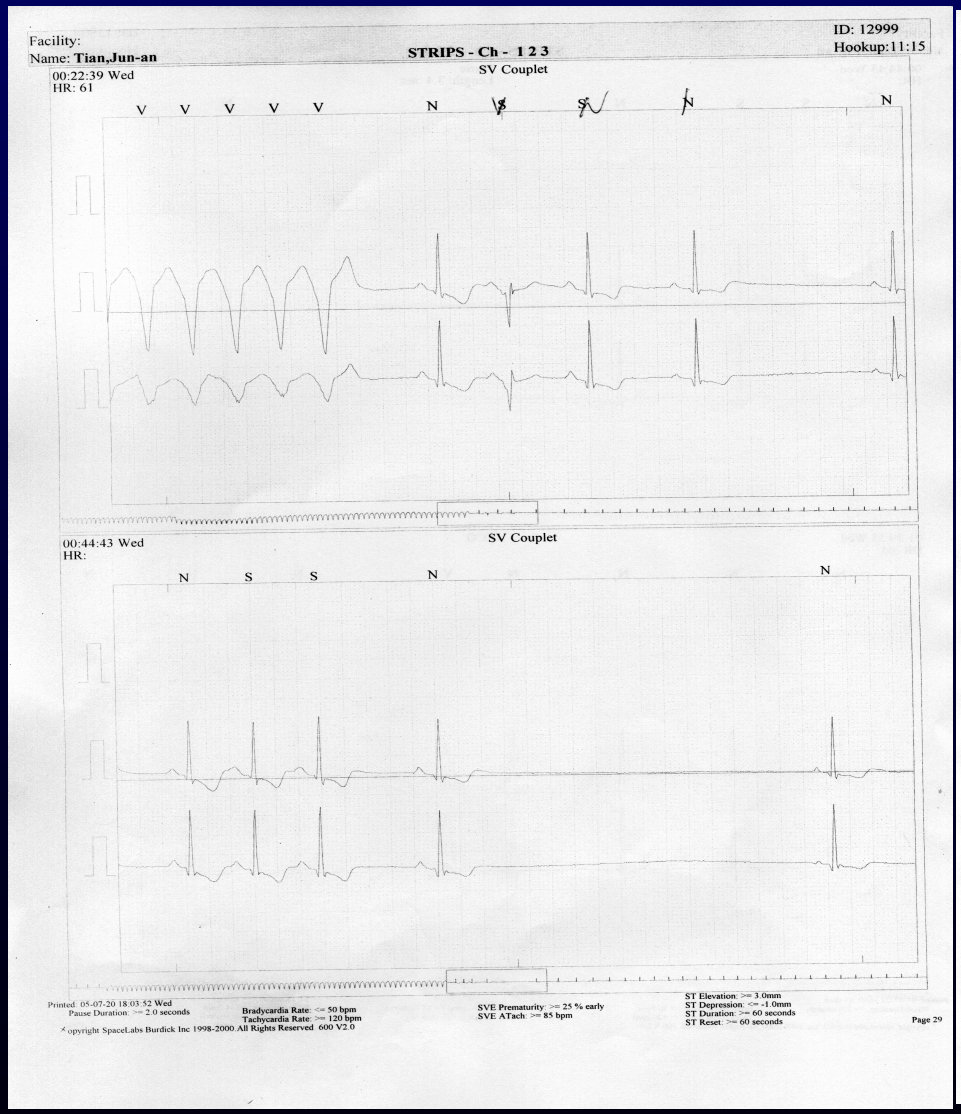


Fig 6　Holter：sinus rhythm, sinus arrest (3.6 s)

Sustained VT (last 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 drug
- 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
- Sometime her EKG showed PVC
- Aspirin 100mg qd

(Living in the countryside, regularly monitor INR is difficult for her, so, no anticoagulation)

Discuss

- **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 ventricular 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 hightly 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 potential usefulness for H_2O_2 imaging as a supplement to 2D UCG in the assessment of noncompaction
- It is challenge for us how to treat this patient.

  For sinus arrest, we thought it may be related with propafenone, we just stopped this drug, and follow-up.

  For VT, we chose the ablation therapy

Question

What is the optimal therapy for this case?