

Isolated Noncompaction of Right Ventricular Myocardium and Arrhythmia

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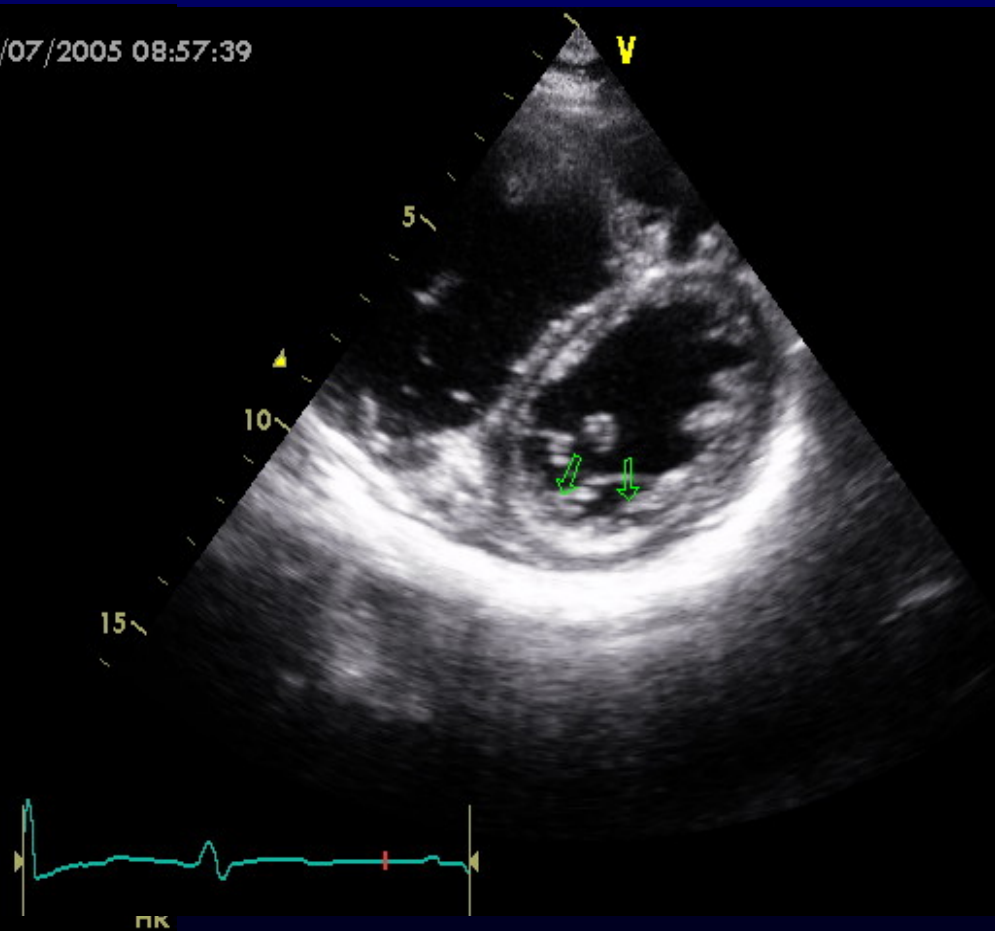
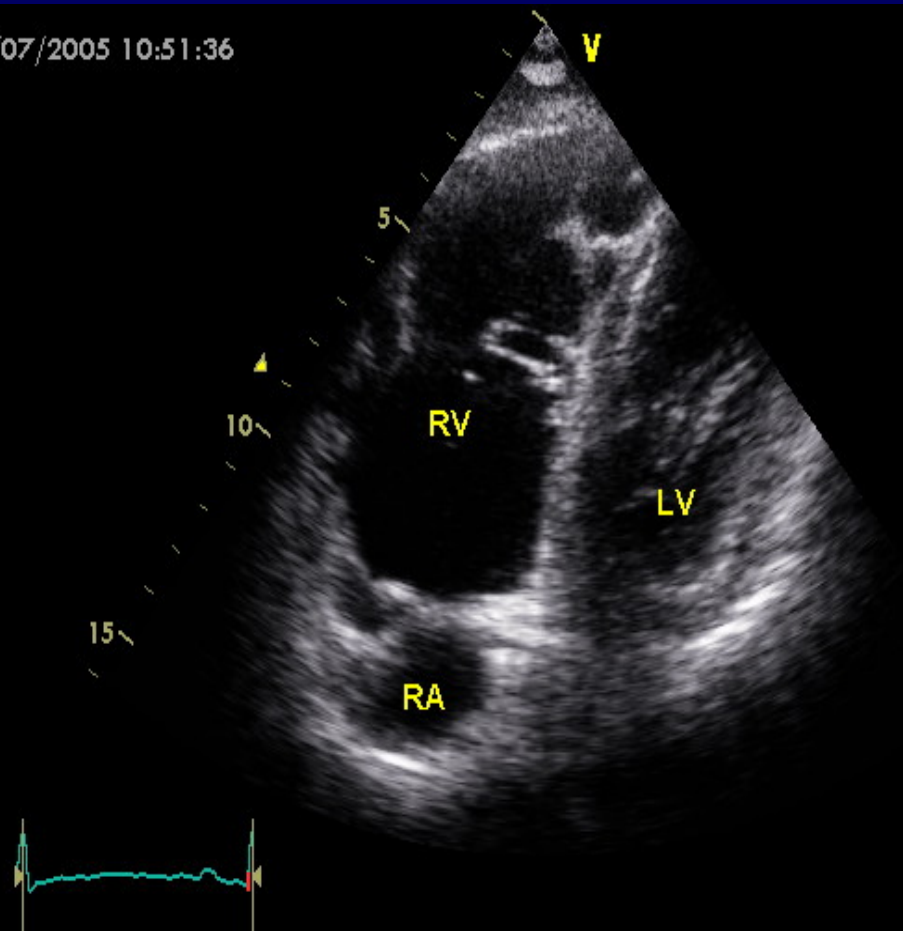
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- **A 28 years-old woman**
- **Palpitation, chest distress for 2 months**
- **Had a syncope history with ventricular tachycardia**
- **No embolism event**
- **Propafenone: oral dose 150mg tid for 2 months**
- **No family history of cardiomyopathy**

- **HR: 64 b/m, BP: 94/64mmHg**
- **Jugular venous pressure was normal**
- **No cardiac murmur**
- **A chest X ray showed marked cardiomegaly**
- **EKG: nonspecific 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**

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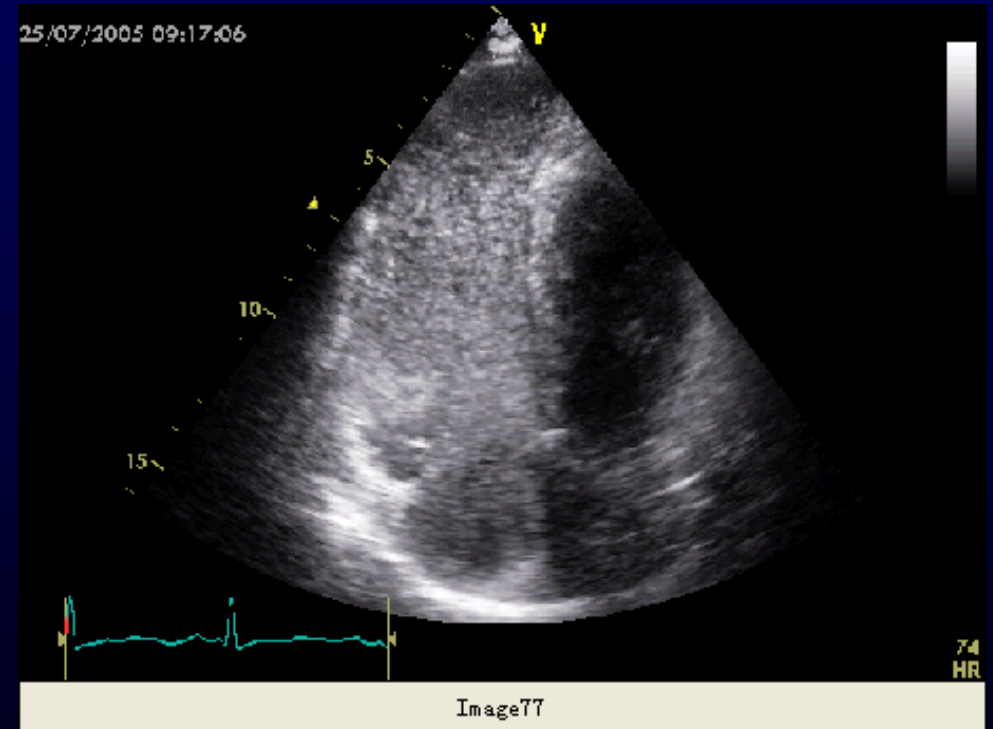
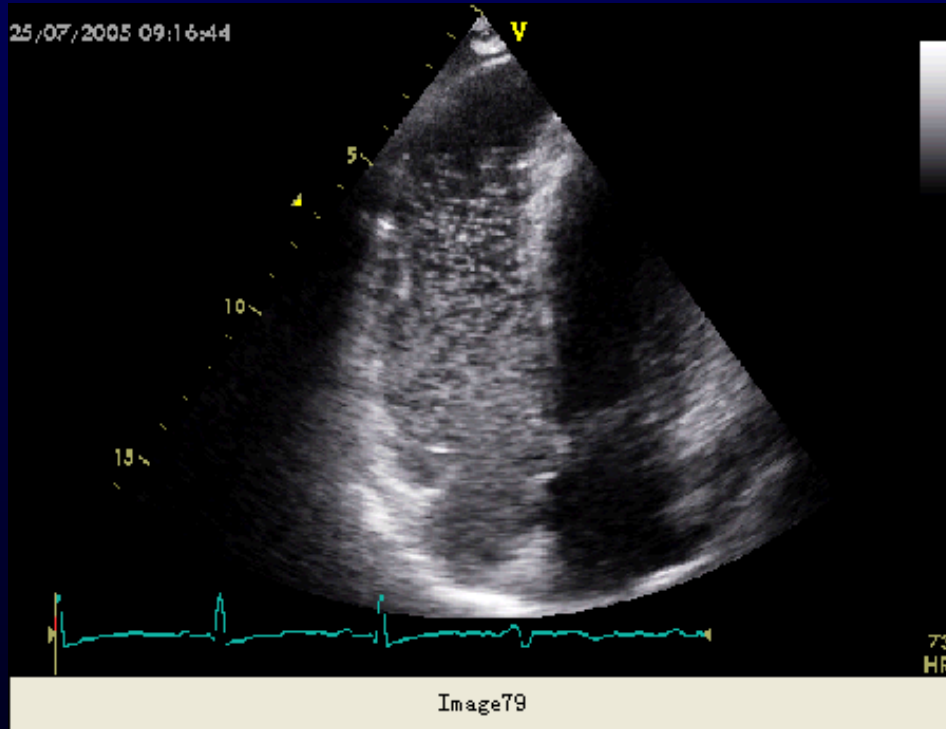
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A: the 4-chamber view

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

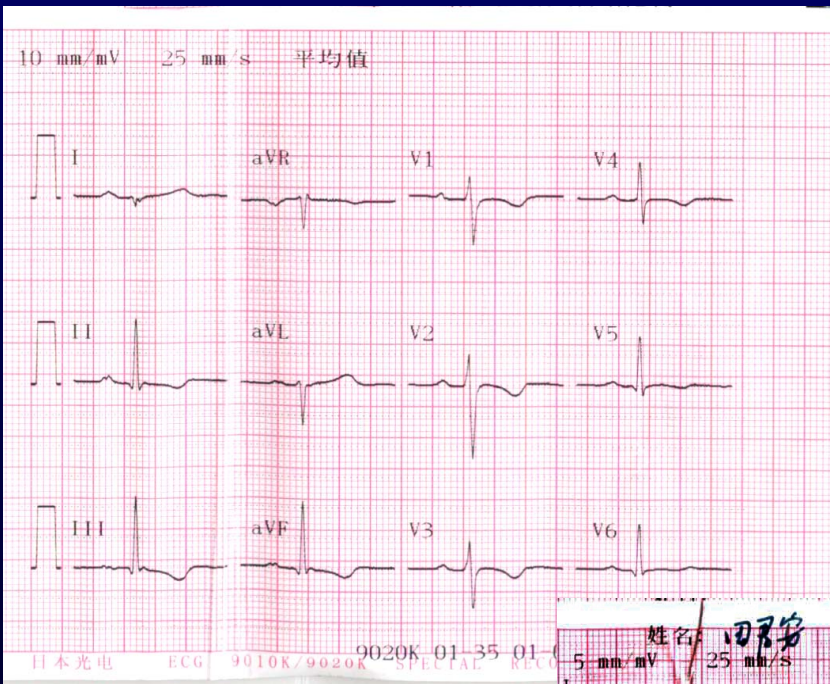
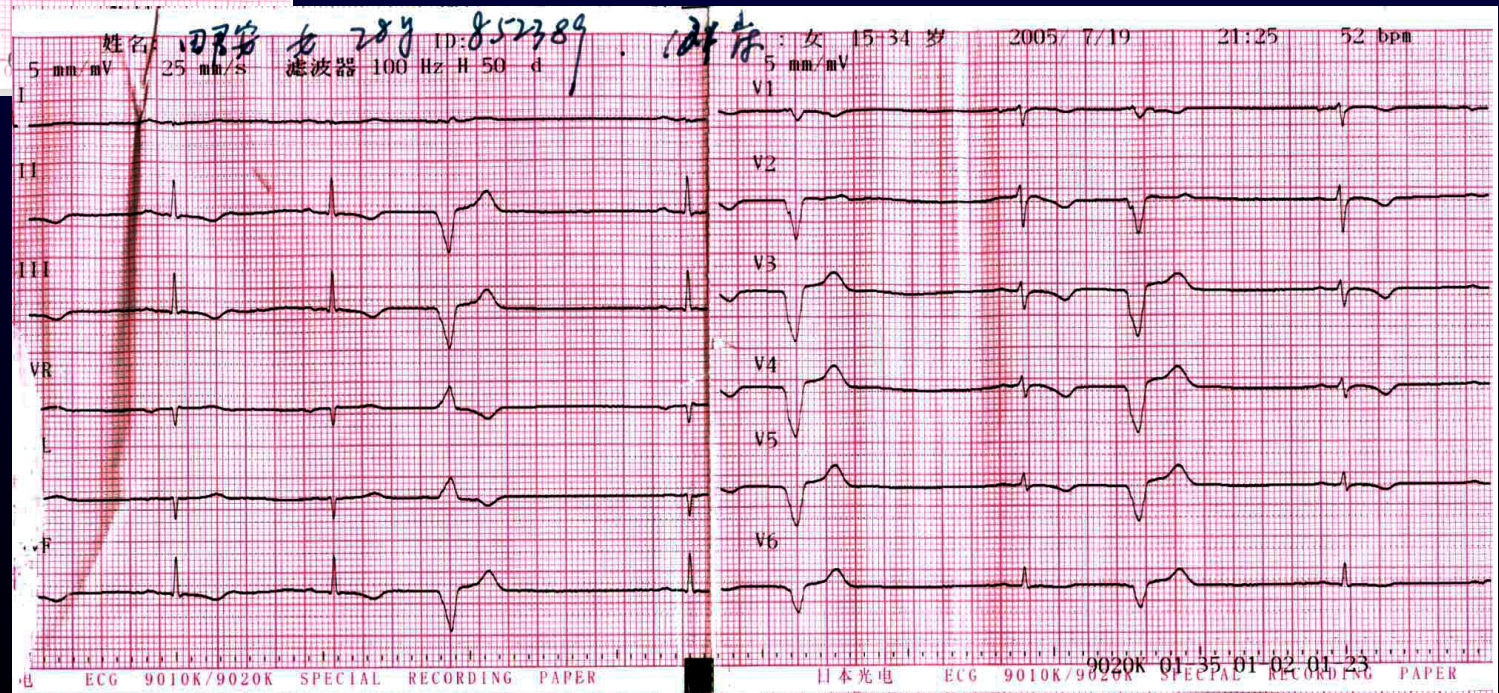


Fig 4 The resting EKG showing repolarization changes, inverted T wave, ST segment changes and premature ventricular complexes



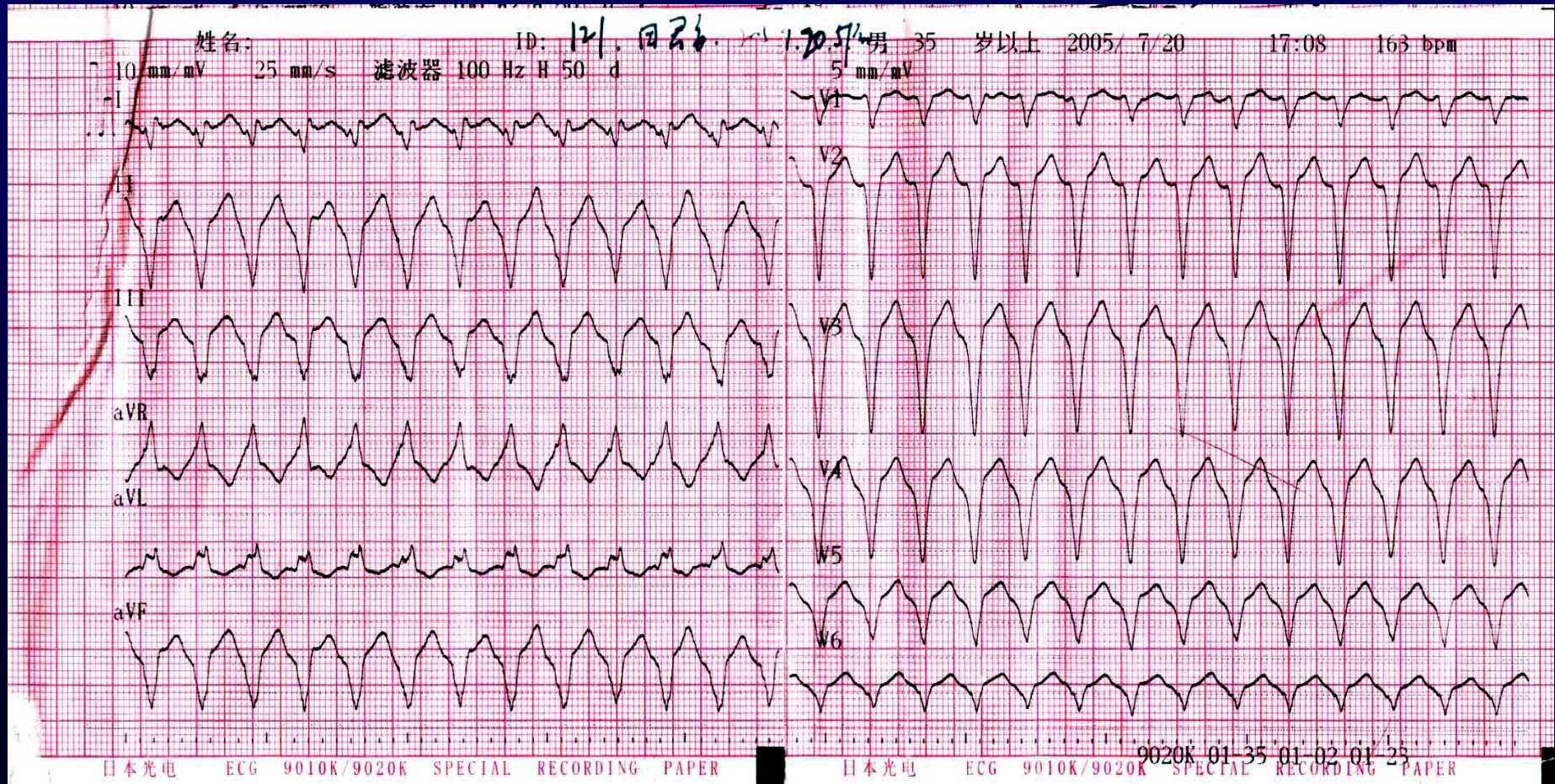
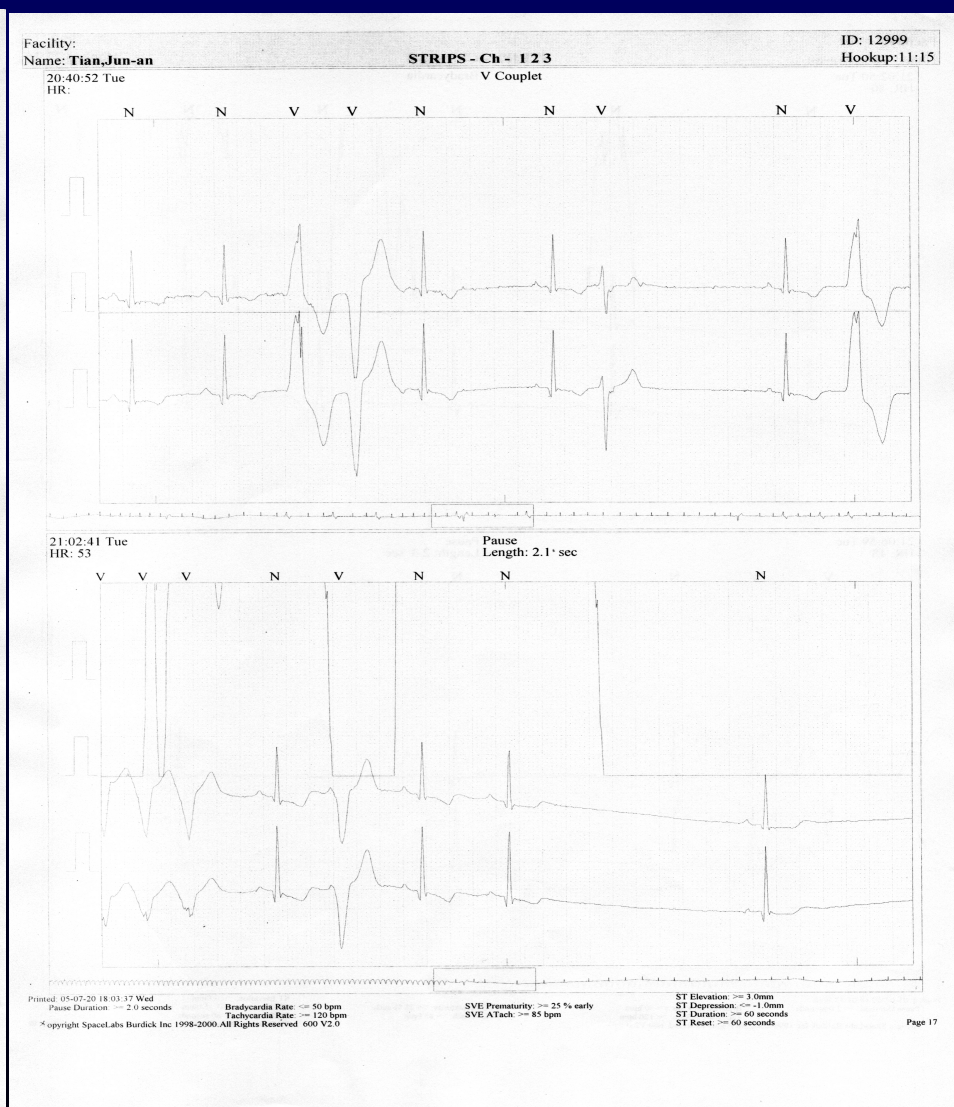
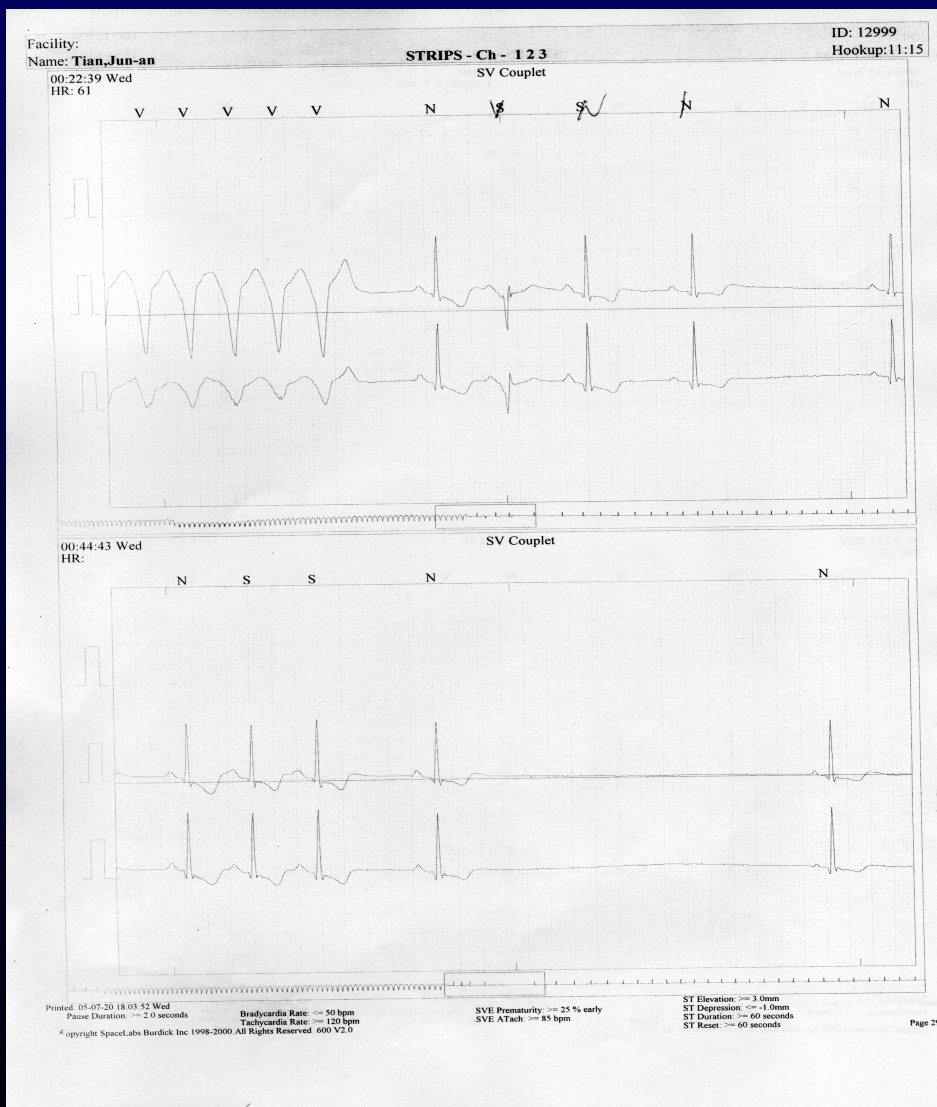


Fig 5 Symptoms (dizziness、amaurosis and a blurred vision) occurring during sustained ventricular tachycardia. Stop VT after IV administration Amiodarone



**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 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 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?