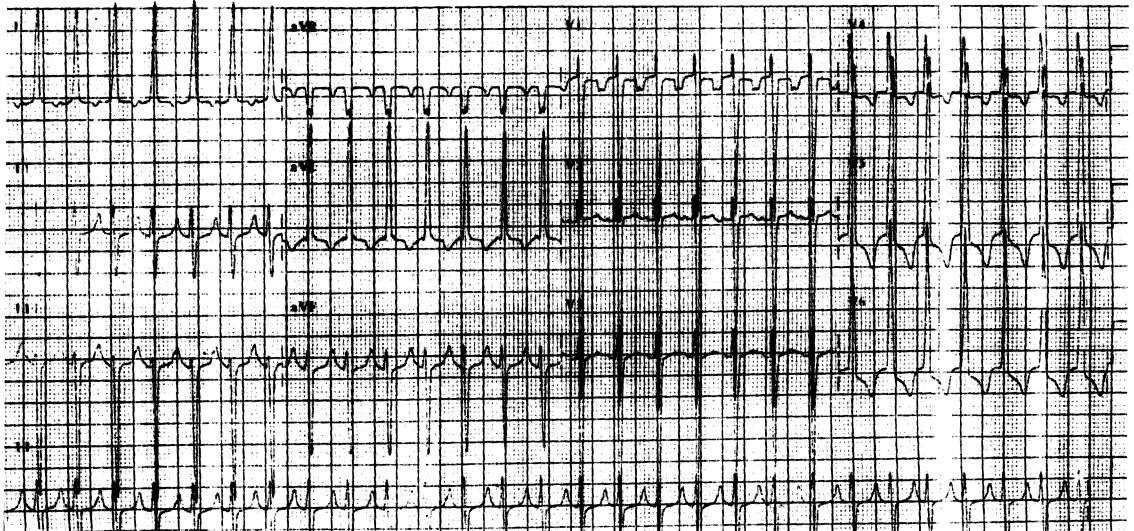


Cardiac Dysrrhthmia in Systemic Disease

S C Quek, K S Ng, P S Low



ABSTRACT

This is an ECG of a 6-year-old girl. She presented to us for managment of epilepsy. Magnetic resonance imaging (MRI) of the brain showed multiple lesions consistent with cortical and subcortical tubers. There were also achromic spots on her skin and echocardiography demonstrated a rhabdomyoma near the right ventricular outflow tract. A diagnosis of tuberous sclerosis, an autosomal-dominantly inherited condition, was made. What is abnormal about the ECG?

Division of Cardiology
Department of Paediatrics
National University Hospital
5 Lower Kent Ridge Road
Singapore 119074

S C Quek, MBBS, FAMS
Senior Lecturer and Consultant

P S Low, MD, FAMS
Chief

Cardiac Department
National University Hospital

K S Ng, MBBS, FAMS
Consultant

Correspondence to:
Dr S C Quek

DIAGNOSIS

Supraventricular tachycardia
Left ventricular hypertrophy

DISCUSSION

In this ECG, there is a tachycardia of 169 beats per minute. The QRS complexes are normal. It is difficult to define normal p waves on this ECG, and a clinical diagnosis of supraventricular tachycardia would be appropriate. The subtype and mechanism of supraventricular tachycardia is best ascertained after more sophisticated investigation such as an electrophysiologic study.

There is also evidence of left ventricular hypertrophy suggested by the tall R in V5, 6 and deep S in the corresponding right precordial leads. The chest X-ray showed cardiomegaly, and the left ventricular hypertrophy and dilation are confirmed on M-mode measurements on the echocardiogram. This heart enlargement is most likely the result of the long-standing tachyarrhythmia.

The patient fulfills the clinical criteria for the diagnosis of tuberous sclerosis⁽¹⁾. This disorder is known to be associated with primary cardiac tumours⁽²⁾. Histologically, these are benign rhabdomyomas⁽³⁾ which may spontaneously regress with time. A conservative approach to the management⁽⁵⁾ of these tumours can frequently be

adopted, unless causing significant haemodynamic compromise.

Cardiac arrhythmias⁽⁴⁾ have also been described. The long-term outcome is uncertain, but supportive and anti-arrhythmic therapy may be necessary. Given the background of tuberous sclerosis, it is plausible that a tuber might be a focus of automaticity, or act as a substrate for re-entrant of tachycardia. It seems reasonable, therefore, to hope for resolution of the rhythm disorder with improvement in ventricular function following spontaneous regression of cardiac tuber.

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