His bundle tachycardia

Author: Doctor Elisabeth Villain
Creation Date: January 2003

Scientific Editor: Doctor Damien Bonnet

1Service de cardiologie pédiatrique, Hôpital Necker - Enfants Malades, 149 Rue de Sèvres, 75743 Paris Cedex 15, France. elisabeth.villain@nck.ap-hop-paris.fr

Abstract
Congenital His bundle tachycardia or junctional ectopic tachycardia (JET) is a rare arrhythmia observed in neonates and infants less than 6 months. Tachycardia is due to increased automaticity within the atrioventricular node and the His bundle but its etiology remains unknown, although familial cases are frequent. The typical electrocardiogram of JET is characterized by a narrow QRS tachycardia with atrioventricular dissociation and occasional sinus capture beats. The ventricular rate usually ranges from 150 to 350 beats per minute (bpm). JET is a severe arrhythmia causing tachycardia-induced cardiomyopathy and death when it is undiagnosed or uncontrolled. It is resistant to conventional antiarrhythmic drugs and the most effective antiarrhythmic agent is oral amiodarone which decreases the rate of JET, resulting in suppression of symptoms and normalization of echocardiography. The treatment has to be maintained until the sinus rhythm becomes faster than the ectopic rhythm, which may take years. Radiofrequency ablation of the His bundle has been proposed in patients who are refractory to antiarrhythmic drugs, but it carries a high risk of atrioventricular block. Long-term prognosis of patients with JET is unknown but most patients of those who stopped medical treatment are doing well.

Keywords
Supraventricular tachycardia, His bundle tachycardia, junctional ectopic tachycardia, amiodarone, neonatal arrhythmia.

Disease name and synonyms
Congenital His bundle tachycardia, congenital junctional ectopic tachycardia (JET)

Excluded diseases
Post-operative junctional ectopic tachycardia

Definition
Congenital JET was first described as a distinct entity by Coumel and coworkers in 1875. JET is thought to be congenital since it was noted in infants less than 6 months. The typical electrocardiogram (ECG) of JET is characterized by narrow QRS tachycardia and atrioventricular (AV) dissociation, with sinus P wave wandering...
through the QRS at a rate slower than the junctional rate; there may be occasional sinus capture beats. JET with retrograde conduction has also been described.

Differential diagnosis
JET with retrograde ventriculoatrial conduction may be difficult to differentiate from a supra-ventricular reciprocating tachycardia. In JET with retrograde conduction, intravenous adenosine or adenosine triphosphate will slow down ventriculoatrial conduction, without changing the ventricular rate; in reciprocating tachycardia, these drugs have no effect on tachycardia or convert it to sinus rhythm.

Clinical description
JET is a malignant arrhythmia causing tachycardia-induced cardiomyopathy and sudden death when it is undiagnosed or uncontrolled. Patients present with a varying degree of congestive heart failure and a rapid heart rate. In a multicenter study including 26 infants with JET, the heart rate was reported to range from 150 to 350 beats per minute (bpm) with a mean of 230 bpm. The heart structure of these patients is normal, with a dilated left ventricle and reduced shortening fraction at echocardiography.

Management
After birth, ventilatory support and treatment of cardiac failure are often necessary. JET is known for its resistance to conventional antiarrhythmic drugs and its potentially poor prognosis. Treatment is indicated in children with rapid heart rate symptoms and reduced cardiac function. The most effective antiarrhythmic agent is amiodarone, that should be given orally, with a loading dose of 500 mg/m2 during eight days followed by a maintenance dose of 250 mg/m2. Amiodarone results in slowing down tachycardia, with a significant decrease of the ventricular rate within the first days of treatment. The treatment has to be maintained until the sinus rhythm becomes faster than the ectopic rhythm, which may take several years. Successful treatment of JET with oral sotalol, propafenone and flecainide has also been reported.

Radiofrequency ablation of the His bundle has been proposed in patients refractory to antiarrhythmic drugs, including amiodarone. Risk of atrio-ventricular block is high.

Etiology
JET is due to increased automaticity within the atrioventricular node and the His bundle but its etiology remains unknown. JET evolving into complete heart block has been described. Very little correlative pathoanatomical research has been conducted. Multifocal Purkinje cell tumors have been found in the conduction system of a 13-month-old boy with JET who died of cardiac arrest. Histological studies have shown congenital abnormalities of the AV junction and left-sided AV bundle.

Diagnostic methods
Surface ECG is usually sufficient for diagnosis of JET. Intracardiac recordings show that JET originates in the atrioventricular junction, with a His bundle potential preceding each ventricular depolarization with a normal HV interval. The mechanism of JET is determined to be an automatic focus, as rapid atrial or ventricular pacing may temporarily overdrive the tachycardia, which resumes immediately after cessation of pacing. Cardioversion does not stop tachycardia.

Genetic counseling
JET was found to be familial in half of the 26 cases reported by Villain et al. No gene has been identified yet.

Antenatal diagnosis
Cases of JET have been characterized and treated in utero in patients with antenatal tachycardia and cardiac failure. When JET is diagnosed before birth, intrauterine management should aim at maintaining adequate cardiac output to postpone the delivery until lung maturation is achieved. Treatment with sotalol and amiodarone has been reported to slow down the ventricular rate of fetuses with JET.

Unresolved questions
Etiology of JET remains unknown, as well as the late prognosis of patients who may have unrecognized His bundle lesions.

References
Janousek J, Paul T. Safety of oral propafenone in the treatment of arrhythmias in infants and


