CASE STUDY

Junctional ectopic tachycardia in six paediatric patients

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Abstract

The presenting features and treatment responses of six children with junctional ectopic tachycardia are evaluated. Two of the patients were siblings and both presented in early childhood with cardiopulmonary failure. The elder sibling died, the surviving sibling was controlled on a combination of amiodarone, digoxin, and sotalol. The remaining four patients presented in later childhood with tachycardia induced cardiomyopathy. Two of the patients were diagnosed incidentally and have normalised their myocardial function on sotalol therapy. The other two presented in congestive cardiac failure. Radiofrequency His bundle ablation and insertion of a permanent pacemaker to control the arrhythmia was undertaken in the elder of the two patients. The remaining patient has had marginal recovery of myocardial function on a combination of amiodarone and sotalol treatment. Improvement in myocardial function may take several months and is dependent on control of the tachycardia in some patients. Sotalol, when used as single or combination treatment, was partially successful in four cases in reducing heart rate. None of the patients reverted to sinus rhythm.

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Junctional ectopic tachycardia (JET) is a rare tachyarrhythmia peculiar to paediatric cardiology and is recognisable on surface electrocar-

diogram by a narrow QRS tachycardia with atrioventricular dissociation and a faster ventricular rate.¹ The origin of the tachycardia is an automatic ectopic focus in the nodal conduction tissue above the bifurcation of the bundle of His.² The arrhythmia is classified as congenital when diagnosed in patients younger than 6 months; a family history has been observed in 50% of patients with this form of JET.³ It may be found in association with congenital heart defects—for example, atrial and ventricular septal defects.¹ Acquired JET may occur postoperatively particularly in tetralogy of Fallot and ventricular septal defect repairs.⁴ Despite advances in pharmacological treatment, 35% of patients die as a result of congenital JET. His bundle ablation and insertion of a pacemaker may be a lifesaving measure in patients with severe congestive heart failure not responsive to medical therapy.³

Patients and methods

A retrospective evaluation of the presenting features and treatment outcome of six children with a diagnosis of JET was carried out at two institutions between 1986 and 1994. None of the patients had underlying congenital heart disease and none had undergone intracardiac surgery. The diagnosis of JET was made electrocardiographically according to established criteria. An example of a typical electrocardiograph taken from patient 3 (table 1) is shown in fig 1. Echocardiographic assessment of myocardial contractility was carried out in four patients at initial assessment. Cardiac enzymes and thyroid function tests were performed on all but one patient in an attempt to exclude the presence of acute myocarditis and thyrotoxicosis, respectively. None of the patients underwent myocardial biopsy.

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Table 1 Presenting features and treatment responses

Case No	Age	Sex	FH	CCF	AR (per min)	VR (per min)	SF	Failed treatment	Successful treatment	Treatment response
1	6 weeks	F	+	Severe	158	260	Poor	S, D	A, D, S	Normal function, VR 120/min
2	12 weeks	M	+	Severe	142	272	Poor	Ver + CV	–	Died
3	10 months	F	–	Moderate	150	214	7%	D	A, S	SF 18%, VR 100/min
4	4 years	F	–	–	100	140	25%	–	S	Normal function, VR 74/min
5	7 years	M	–	–	90	180	25%	–	S	Normal function, VR 120/min
6	8 years	M	–	Shock	130	230	3%	S, A	Pacemaker	SF 10%, VR 80/min

A, amiodarone; AR, atrial rate; CCF, congestive cardiac failure; CV, cardioversion; D, digoxin; FH, family history; S, sotalol; SF, shortening fraction; Ver, verapamil; VR, ventricular rate; –, absent; +, present.

Figure 1 Lead 11 surface electrocardiograph strip from patient 3 at 4 years old, on sotalol and amiodarone. A narrow QRS complex tachycardia with a dissociated slower atrial rate is evident. Paper speed, 25 mm/sec. R P waves.

Results

The presenting features and treatment responses of the six patients are summarised in table 1. None of the patients reverted to sinus rhythm and five are alive after follow up of 12 months to five years. Two patients were of mixed heritage and four were black. The former two patients were siblings with different fathers. Patient 2 died at the age of 12 weeks from a suspected supraventricular tachycardia shortly after an unsuccessful attempt at cardioversion and intravenous verapamil treatment. A later review of the electrocardiogram confirmed a diagnosis of JET. The sibling presented similarly and has survived to the age of 5 years on a combination of amiodarone (7.5 mg/kg/day), digoxin (3 µg/kg/day), and sotalol (7.5 mg/kg/day). Her cardiac function normalised within 24 months of treatment. Attempts at withdrawal of sotalol resulted in recurrence of the tachycardia. An ophthalmological assessment after being on amiodarone for three years revealed bilateral corneal deposits below the visual axis. These two patients have been the subjects of a previous case report.⁵

The remaining four children were much older at presentation. Two patients were completely asymptomatic from their arrhythmias diagnosed incidentally when presenting with unrelated problems. They both had mildly reduced left ventricular function that normalised on sotalol treatment (6 mg/kg/day in patient 4, 15 mg/kg/day in patient 5) over a period of 12 months.

Patient 3 presented with a suspected idiopathic cardiomyopathy and was treated initially with digoxin and diuretics. At 4 years old the incessant tachycardia that accompanied the cardiomyopathy was recognised as JET. The tachycardia was slowed using a combination of sotalol (3 mg/kg/day) and amiodarone (12 mg/kg/day). Her cardiac function improved over 12 months and she remains well controlled on amiodarone (7 mg/kg/day) alone, after the successful withdrawal of sotalol.

The oldest of the six patients presented in cardiogenic shock. Initial success with reduction of his heart rate to 100 beats/min was achieved using amiodarone (12 mg/kg/day) and sotalol (3 mg/kg/day). However, 24 hour electrocardiographic monitoring revealed breakthrough episodes of tachycardia up to 150 beats/min. In addition, a trial of home treatment was unsuccessful because of non-compliance. Radiofrequency His bundle ablation and insertion of a permanent pacemaker

was undertaken. The His bundle could not be located as expected from the right ventricular approach. Successful ablation was eventually carried out from the left ventricular approach, and marginal recovery of ventricular function has occurred during the subsequent two year follow up of the patient.

Discussion

AGE AND MODE OF PRESENTATION

Most cases of JET described in the literature are thought to be congenital and present before the age of 6 months.³ Four of the patients in this series presented at a much older age (10 months to 8 years). Adult patients with JET have been reported, two of whom had underlying congenital heart defects (atrial septal defect and a ventricular septal defect).⁶ Another patient presented with JET during a febrile illness.⁷ The onset of JET in our patients was not preceded by any significant clinical event such as febrile illness, and none of the patients had associated cardiac defects.

The later presentation of the study patients may be related to poor access to primary health care facilities and socioeconomic factors.

DRUG TREATMENT

JET is very difficult to treat medically and is unresponsive to synchronised cardioversion.¹ Spontaneous cessation of the tachycardia may occur, therefore temporary antiarrhythmic treatment is the treatment of choice.³ Adequate control of the tachycardia resulted in improvement of myocardial function in four of the patients in our case series. Patients with the worst left ventricular function had faster ventricular rates, whereas those with a more benign course had slower rates. The presentation of JET with poor left ventricular function in the majority of the patients emphasises the importance of excluding a tachyarrhythmia as a cause for a myopathic heart.

Amiodarone is reportedly one of the more effective agents used to treat patients with both congenital and postoperative forms of JET,^{3 8 9} but its alleged side effects limit its chronic use.¹⁰ Fortunately, toxic effects appear to be rare in children younger than 10 years old.¹¹ Corneal microdeposits (found in patient 1) is one of the more frequent side effects, but is usually asymptomatic.¹² The recommended oral loading dose is 10 mg/kg for 10 days followed by a maintenance dose of 5 mg/kg in children.¹³

Combination drug therapy was used in three patients to avoid the use of prolonged high dose amiodarone therapy. Two of the study patients with mildly reduced left ventricular function were successfully and exclusively treated with sotalol. It has been used successfully to treat one other patient with JET in the literature. The dose prescribed by the authors in the treatment of various arrhythmias is 2–8 mg/kg/day.¹⁴ As with any β adrenergic antagonist, careful dose titration is essential in patients with reduced cardiac performance.¹⁵

Although digoxin has a poor effect on the tachycardia in JET and may induce arrhythmias such as ventricular fibrillation and atrial

tachycardia, it has aided in controlling congestive cardiac failure in patients with JET.³ Amiodarone increases plasma digoxin concentrations, therefore a reduction in the dose of digoxin is required if used as adjunct therapy.¹⁶ A small dose of digoxin was used in patient 1.

PACING

The difficulty in locating the His bundle from the right sided approach during electrophysiological studies in patients with JET has been noted by other investigators. Villain *et al* suggest these difficulties may be encountered because either the His bundle is left sided in patients with congenital JET or JET is a ventricular tachycardia originating from the left side of the ventricular septum.³ However, it is possible that technical difficulties may have prevented the detection of the His bundle potential from the conventional right sided approach in our patient.

In conclusion, in our experience JET may present in older children without a family history or a preceding historical event. It may masquerade initially as a supraventricular tachycardia. Various degrees of severity may occur and if incessant the tachycardia may result in severe left ventricular dysfunction. Although amiodarone is reportedly the antiarrhythmic agent of choice, sotalol as the sole drug was successfully used to control the tachycardia in two study patients with a more benign form of JET.

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