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Evaluation of a Staged Treatment Protocol for Rapid Automatic Junctional Tachycardia After Operation for Congenital Heart Disease

EDWARD P. WALSH, MD, FACC, J. PHILIP SAUL, MD, FACC,
GARY F. SHOLLER, MB, BS, FACC, JOHN K. TRIEDMAN, MD, FACC,
RICHARD A. JONAS, MD, FACC, JOHN E. MAYER, MD, FACC, DAVID L. WESSEL, MD

Boston, Massachusetts

Objectives. **This study sought to 1) develop an efficient treatment protocol for postoperative automatic junctional tachycardia (JT) using conventional drugs and techniques, and 2) identify clinical features associated with this disorder by analyzing a large study group.**

Background. **Postoperative JT is a transient arrhythmia that may be fatal after operation for congenital cardiac defects. Its precise cause is unknown. A variety of palliative treatments have evolved, but because of a low incidence of JT, large studies of the most efficient therapeutic sequence are lacking.**

Methods. **A protocol for rapid JT (>170 beats/min) was adopted in 1986, and was tested in 71 children between 1986 and 1994. Staged therapy involved 1) a reduction of catecholamines; 2) correction of fever; 3) atrial pacing to restore synchrony; 4) digoxin; 5) phenytoin or propranolol or verapamil; 6) procainamide or hypothermia; and 7) combined procainamide and hypothermia. Effective therapy was defined as a sustained reduction of JT rate <170 beats/min within 2 h. Clinical profiles of the study group were contrasted with all patients without JT from this same era to identify features associated with JT.**

Results. **Of the multiple treatment stages, only correction of fever and combined procainamide and hypothermia appeared to be efficacious. By refining the protocol to eliminate nonproductive stages, the time to JT control was significantly shortened for the last 30 patients. Treatment was ultimately successful in 70 of 71 children. Postoperative JT was strongly associated with young age, transient atrioventricular block and operations involving ventricular septal defect closure.**

Conclusions. **A staged approach to therapy, with emphasis on combined hypothermia and procainamide in difficult cases, appears to be an effective management strategy for postoperative JT. These results may also serve as comparison data for evaluation of newer and promising JT options, such as intravenous amiodarone. Trauma to conduction tissue may play a central role in the etiology of this disorder.**

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Junctional tachycardia (JT) is an uncommon arrhythmia thought to arise from abnormal automaticity at the atrioventricular (AV) node or proximal His bundle (1,2). In children, it may occur as a primary idiopathic disorder (3) but is seen more often as a transient phenomenon immediately after surgery for congenital heart defects (1,4). Although mildly elevated junctional rates are well tolerated in the postoperative setting, rapid JT can result in hemodynamic compromise or death, and remains among the more difficult tachycardias to control. Fortunately, it will usually resolve spontaneously within hours or days if the patient can be supported by restoring AV synchrony and slowing the junctional rate to a physiologic range. A variety of interventions have evolved to accomplish these goals, including pacing maneuvers (5,6), pharmacologic therapy (7–11), induced hypothermia (12–14) and even His bundle ablation (15). With this array of options, postoperative JT can now be managed successfully in most patients, but systematic study of the most efficient treatment sequence has not been performed in a large group. This report expands significantly the clinical experience with postoperative JT by describing results of a staged therapeutic protocol used in 71 consecutive patients at Children's Hospital in Boston from 1986 to 1994.

Methods

Protocol development. A management plan for rapid JT was designed in 1986 (Fig. 1A) based on published recommendations (4,7,12) and institutional experience (16). Periodic modifications were made to the original protocol as preliminary data were reviewed, leading to a refined scheme (Fig. 1B) that was adopted in 1990 and remained in effect until study completion in 1994. Intravenous propafenone was not available at the hospital during this period, and intravenous amio-

From the Departments of Cardiology and Cardiovascular Surgery, Children's Hospital, and Departments of Pediatrics and Surgery, Harvard Medical School, Boston, Massachusetts. This work was supported by the Sean Roy Johnson Memorial Heart Fund, Boston Children's Hospital.

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Address for correspondence: Dr. Edward P. Walsh, Department of Cardiology, Children's Hospital, 300 Longwood Avenue, Boston, Massachusetts 02115. E-mail: epwalsh@cardio.tch.harvard.edu.

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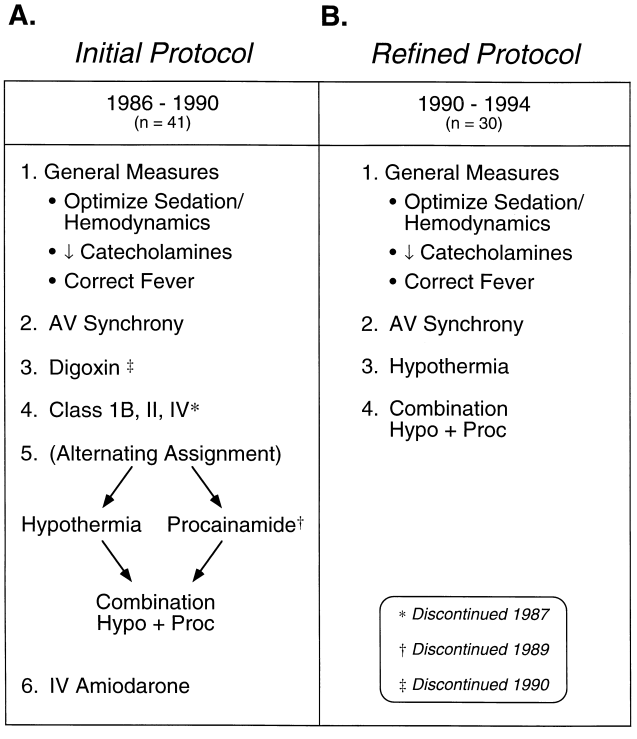
Abbreviations and Acronyms

AV = atrioventricular
ECG = electrocardiogram
JT = automatic junctional tachycardia
TOF = tetralogy of Fallot
VSD = ventricular septal defect

darone was used in just one patient under a compassionate use protocol.

Patient selection and diagnosis of JT. The diagnosis was based on established electrocardiographic (ECG) criteria (1), including 1) QRS configuration similar to conducted sinus or atrial paced beats; 2) rapid ventricular rate greater than or equal to atrial rate, usually exhibiting a pattern of "warm up" at initiation; and 3) atrial activity in the form of dissociated sinus rhythm or retrograde conduction in patterns of 1:1 or Wenckebach. If atrial timing was indistinct on the surface ECG, electrograms were recorded from pacing wires or an esophageal lead. In cases where the A/V ratio was 1:1 or tachycardia onset seemed abrupt, or both, attempts were made to exclude a reentrant mechanism with atrial pacing maneuvers, intravenous adenosine or electrical cardioversion. Abrupt tachycardia termination in response to any of these measures was considered incompatible with a diagnosis of automatic JT, and such patients were excluded.

Figure 1. Outline of initial protocol **(A)** and refined protocol **(B)** for management of rapid postoperative JT, used from 1986 to 1994. (See text for details.) Hypo = hypothermia; IV = intravenous; Proc = procainamide.



Patients with a confirmed diagnosis were included in this study only if JT was faster than 170 beats/min. This value was chosen based on our institutional experience, which suggested that JT begins to exert deleterious hemodynamic effects at or above this rate. Patients with slower JT were excluded, as were those already receiving a class I or class III antiarrhythmic drug for any condition.

The study group included 71 patients. The initial 41, managed from 1986 to 1990, were followed prospectively from the time of JT diagnosis until hospital discharge. Data from these patients were used to generate protocol modifications, which led to the final treatment plan adopted in 1990. Information from an additional 30 consecutive patients treated according to the final protocol from 1990 to 1994 was obtained by retrospective chart review. Clinical features of these 71 patients with JT were then compared with all cardiac surgical patients without JT from this same era. Data on the group without JT were obtained from departmental computer records.

Treatment protocol (Fig. 1). Therapy began with general efforts to optimize hemodynamic variables, respiratory status, electrolyte status and sedation. Attempts were also made to discontinue catecholamine infusions whenever the hemodynamic status allowed. In addition, any patient with a temperature >38°C was treated with acetaminophen or a cooling blanket to establish normothermia.

If rapid JT persisted, therapy was begun to restore AV synchrony and slow the junctional rate. Synchrony was achieved with atrial pacing at a rate slightly faster than JT using epicardial wires or an esophageal lead. The pacemaker rate was manually adjusted at frequent intervals to maintain the minimal rate for effective synchrony. Atrial pacing was not used as isolated therapy unless (or until) the JT rate was <200 beats/min.

Direct therapy to slow the junctional rate with drugs or hypothermia was administered promptly to all patients with JT >200 beats/min or to any patient with persistently elevated rates (170 to 200 beats/min) after a trial of atrial pacing.

Digoxin. For patients not receiving digoxin preoperatively, half of a total digitalizing dose was administered intravenously, with full digitalization over the next 8 to 24 h. Patients receiving the drug preoperatively were given intravenous maintenance doses of 5 μg/kg every 12 h. Digoxin was eliminated from the protocol in 1990 after review suggested minimal efficacy.

Class IB (phenytoin) class II (propranolol) or class IV (verapamil). These agents were tested only during the early months of the protocol (10 trials in six patients). Propranolol and verapamil were restricted to nonhypotensive patients, and verapamil was further restricted to patients over 1 year of age. Intravenous dosages involved phenytoin (10 mg/kg body weight over 20 min), propranolol (0.1 mg/kg over 20 min) and verapamil (0.05 to 0.1 mg/kg over 1 to 5 min). These drugs were abandoned in 1987 because of a lack of efficacy.

Procainamide. Administration began as a bolus of 5 to 15 mg/kg over 15 to 30 min followed by an infusion of 20 to

80 μg/kg per min, aiming for a serum procainamide level of 4 to 10 μg/ml. The infusion was stopped at 12-h intervals to evaluate the necessity for continued use. Procainamide was eliminated as an isolated therapy in 1989 based on an early unfavorable comparison with the results for hypothermia, but was still retained as part of combination therapy.

Hypothermia. Core temperature was reduced to 33 to 35°C using a posterior cooling blanket, with the patient sedated, mechanically ventilated and paralyzed to prevent shivering. Temperature was continuously monitored through a rectal or esophageal thermistor. Patients were allowed to rewarm at 12-h intervals to examine the underlying rhythm. Hypothermia remained an active stage of the protocol for the duration of the trial.

Combined procainamide and hypothermia. Combined therapy involved the drug dosage and cooling technique outlined under *Procainamide* and *Hypothermia*. Combined therapy was retained in the protocol for the duration of the trial.

Amiodarone. Intravenous amiodarone was administered in one patient under a compassionate use protocol with emergency approval of the hospital Committee on Clinical Investigation and parental consent. A single dose of 5 mg/kg was given over 30 min.

Evaluation of treatment response. In an effort to minimize the influence of spontaneous JT resolution on perceived treatment response, *effective* therapy was strictly defined as one which 1) decreased the JT rate <170 beats/min within 2 h, and 2) was not associated with any subsequent rate rebound >170 beats/min. Therapy was defined as *possibly effective* if 1) the time until JT slowing <170 beats/min was delayed beyond 2 h but the patient responded well thereafter and did not require escalation to a higher stage of the protocol, or 2) there were late increases in the JT rate >170 beats/min after at least 6 h of a good early response, whether or not therapy was escalated. *Ineffective* therapy was defined as accelerated or persistently rapid (>170 beats/min) JT requiring escalation of therapy. Two time variables were recorded for each patient: the time to JT control (when sustained rate slowing <170/min was achieved) and the time to JT resolution (when junctional automaticity abated and sinus rhythm returned).

Statistics. Data are presented as mean value ± SD along with median value when appropriate. The chi-square and Fisher exact tests were used to compare categoric variables. Comparison of values for cardiopulmonary bypass time and serum procainamide levels was performed using the unpaired *t* test. For comparisons of time to JT control and time to JT resolution, the log-rank test was performed. A level of $p < 0.05$ was considered significant.

Results

Patient characteristics. Between May 1986 and October 1994, a total of 71 postoperative patients were identified with JT >170 beats/min, representing 1% of all cardiac surgical patients during that period. Ages were 1 day to 18 years (median 4 months, mean 12 ± 28 months). According to the preoperative ECG, all patients were in sinus rhythm, and AV conduction was normal for all but one patient with first-degree block and one with right bundle branch block. Twenty-seven patients (38%) were receiving digoxin preoperatively, and one was receiving propranolol. Cardiopulmonary bypass time was 119 ± 50 min (n = 69), which was not significantly different from the bypass duration of 125 ± 32 min reported by our institution for other large surgical groups (17). Two patients developed JT after procedures that did not involve bypass; both had transposition/intact septum and underwent isolated pulmonary artery banding to prepare the left ventricle for anticipated arterial switch operation.

When the 71 patients with JT were compared with the non-JT group by surgical subcategory (Table 1), there was a

Table 1. Clinical Features of Patients With and Without Automatic Junctional Tachycardia, 1986–1994

	With JT (n = 71) (age 12 ± 28 mo)		Without JT (n = 7,010) (age 48 ± 72 mo)	
Diagnosis	No. of Pts	Mean (±SD) age (% < 1 yr)	No. of Pts	Mean (±SD) age (% < 1 yr)
Positive association				
TOF	26*	6 ± 9 mo (88%*)	910	34 ± 60 mo (53%)
VSD	16*	5 ± 3 mo (94%†)	674	29 ± 56 mo (63%)
TGA/VSD	9†	2 ± 4 mo	198	7 ± 13 mo
Truncus	4†	4 ± 6 mo	90	7 ± 17 mo
No association				
Fontan	6	29 ± 17 mo	585	68 ± 66 mo
TGA/IVS	1	3 mo	381	2 ± 13 mo
CAVC	4	5 ± 1 mo	259	15 ± 30 mo
Caval-PA shunt	—	—	253	45 ± 62 mo
PDA	—	—	238	8 ± 20 mo
Norwood/Stansel	1	1 mo	235	1 ± 4 mo
AO valve	—	—	211	134 ± 102 mo
AV valve	1	5 mo	153	40 ± 60 mo
ASD primum	—	—	151	60 ± 103 mo
PA band	2	42 ± 59 mo	126	10 ± 30 mo
P vein	—	—	112	21 ± 78 mo
P valve/art	—	—	103	52 ± 74 mo
Rastelli	1	32 mo	59	51 ± 54 mo
Transplant	—	—	39	120 ± 106 mo
Negative association				
ASD secundum	—‡	—	421	42 ± 67 mo
Coarct/arch	—‡	—	414	27 ± 53 mo
Pacemaker	—‡	—	391	144 ± 120 mo
Syst-PA shunt	—‡	—	353	19 ± 50 mo
Miscellaneous	—	—	654	64 ± 84 mo

*$p < 0.001$, †$p < 0.01$ and ‡$p = 0.02$ to 0.05 compared with patients without JT. AO = aortic; ASD = atrial septal defect; AV = atrioventricular; Caval-PA shunt = caval to pulmonary shunt (Glenn and variants); CAVC = common atrioventricular canal; Coarct/arch = coarctation and other aortic arch surgery; PA = pulmonary artery; P valve/art = pulmonary valvular or arterial repair or plasty; P vein = total or partial anomalous pulmonary venous return; PDA = patent ductus arteriosus; Syst-PA shunt = systemic to pulmonary arterial shunt; TOF = tetralogy of Fallot and variants; TGA/IVS = atrial (n = 46) or arterial (n = 335) switch for transposition with intact ventricular septum; TGA/VSD = atrial (n = 6) or arterial (n = 192) switch for transposition plus closure of ventricular septal defect; VSD = membranous or muscular ventricular septal defect.

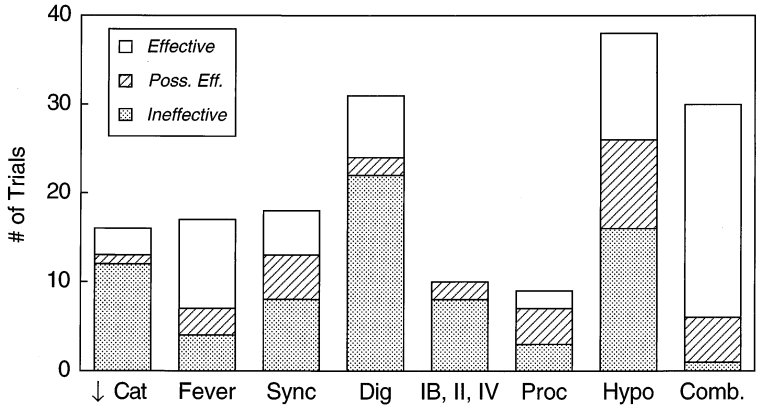


Figure 2. Results of treatment stages for JT in 71 patients. Interventions are arranged (left to right) in general order of testing. Most patients needed to progress through multiple stages of therapy, but an effective (n = 63) or possibly effective (n = 7) treatment was ultimately found in 70 of 71 patients. ↓**Cat** = decrease catecholamine infusion; **Fever** = establishment of normothermia for febrile patients; **Sync** = atrial pacing to restore AV synchrony; **Dig** = digoxin; **IB, II, IV** = class IB (phenytoin [Dilantin]), class II (propranolol) or class IV (verapamil) agent; **Proc** = procainamide; **Hypo** = hypothermia; **Comb.** = combined hypothermia and procainamide; **Poss. Eff.** = possibly effective.

strong association between JT and operations involving ventricular septal defect (VSD) closure, especially tetralogy of Fallot (TOF) repair. For most extracardiac operations (e.g., coarctation) and some intracardiac operations (e.g., atrial septal defect), a negative association was evident. The incidence of JT in the remaining subcategories (e.g., Fontan procedure) did not reach statistical significance.

The mean age at operation (Table 1) was younger for patients with JT compared with patients without it, both overall and within most of the relevant surgical subcategories. In addition, among patients with TOF and VSD (the two surgical subcategories with the majority of JT cases), the proportion of patients under 1 year of age (Table 1) was significantly higher for those with JT compared with those without it.

Although 54 patients (76%) were receiving intravenous catecholamines at the time of JT onset, in most cases the agent was dopamine at a low dose of <5 μg/kg per min. Only 29% were receiving multiple or high dose infusions. Arterial blood gases and serum chemistry data were generally normal at the time of JT onset, with the exceptions of hypokalemia in four patients, hypercapnia in one and metabolic acidosis in one. Correction of these abnormalities did not abolish JT.

A notable postoperative finding was a high incidence of transient AV block, which occurred in 14 of the JT patients (20%). The duration ranged from 1 hour to 10 days, but no patient had permanent block. Rapid JT began at the same time as the return of AV conduction in seven patients, before recovery of conduction in three and sometime after in four. This incidence of transient AV block was significantly higher ($p < 0.001$) than the 2% value observed at our institution for the general cardiac surgical population (18).

Characteristics of JT. Tachycardia typically began in the operating room or soon after arrival in the intensive care unit (median 5 h, mean 21 ± 34 h) but was delayed as late as postoperative day 7 in one child. Peak junctional rates ranged from 170 to 300 beats/min (mean 205 ± 23). The QRS patterns during JT included narrowing (31%), incomplete right bundle branch block (10%), complete right bundle branch block (58%) and bifascicular block (1%). For all but one patient with a prolonged QRS complex during JT, the conduction delay was surgically induced and was a permanent rather than a rate-related aberration. There was complete ventriculoatrial dissociation during tachycardia in 87% of patients, whereas the remaining 13% exhibited retrograde conduction in a 1:1 or Wenckebach pattern.

Hemodynamic variables. The technical/anatomic result of the operation was satisfactory for 85% of the patients with JT. Only 15% had a clinically significant residual hemodynamic defect: dilated systemic ventricle with impaired systolic function (n = 4), moderate or severe AV valve regurgitation (n = 3), elevated right-sided pressures in a Fontan circulation (n = 3) and pulmonary venous obstruction (n = 1).

For 35 patients in whom pressure data were available before and after the onset of JT, systolic arterial blood pressure decreased by a mean of 19 ± 13 mm Hg, and atrial pressures increased by a mean of 4 ± 3 mm Hg in tachycardia. For the remaining patients, the onset of JT was too early in the postoperative course to establish valid comparison data for baseline sinus rhythm. Once JT control was achieved, all patients had arterial and atrial pressures that were physiologically appropriate for age and type of repair.

Results for staged therapy. All 71 patients were treated initially with general supportive measures. The influence of these diverse interventions could not be well quantified, with two exceptions: discontinuation of catecholamine infusions, which was attempted in 16 patients, and establishment of normothermia in 17 patients with fever. Scored results for these and other interventions are shown in Figure 2.

Pacing maneuvers to restore AV synchrony were used as isolated therapy in 18 patients, but were scored as effective in only five. Pacing was also used as adjunctive therapy in an additional 44 patients receiving simultaneous medications or hypothermia, in which case the isolated influence of AV synchrony could not be evaluated.

The initial pharmacologic intervention aimed at slowing JT in the early years of the protocol was digoxin, which was tested in 31 patients and was largely ineffective (Fig. 2). The JT rates for individual patients over the hours after digoxin administration are charted in Figure 3A. Trials of either a class IB, II or

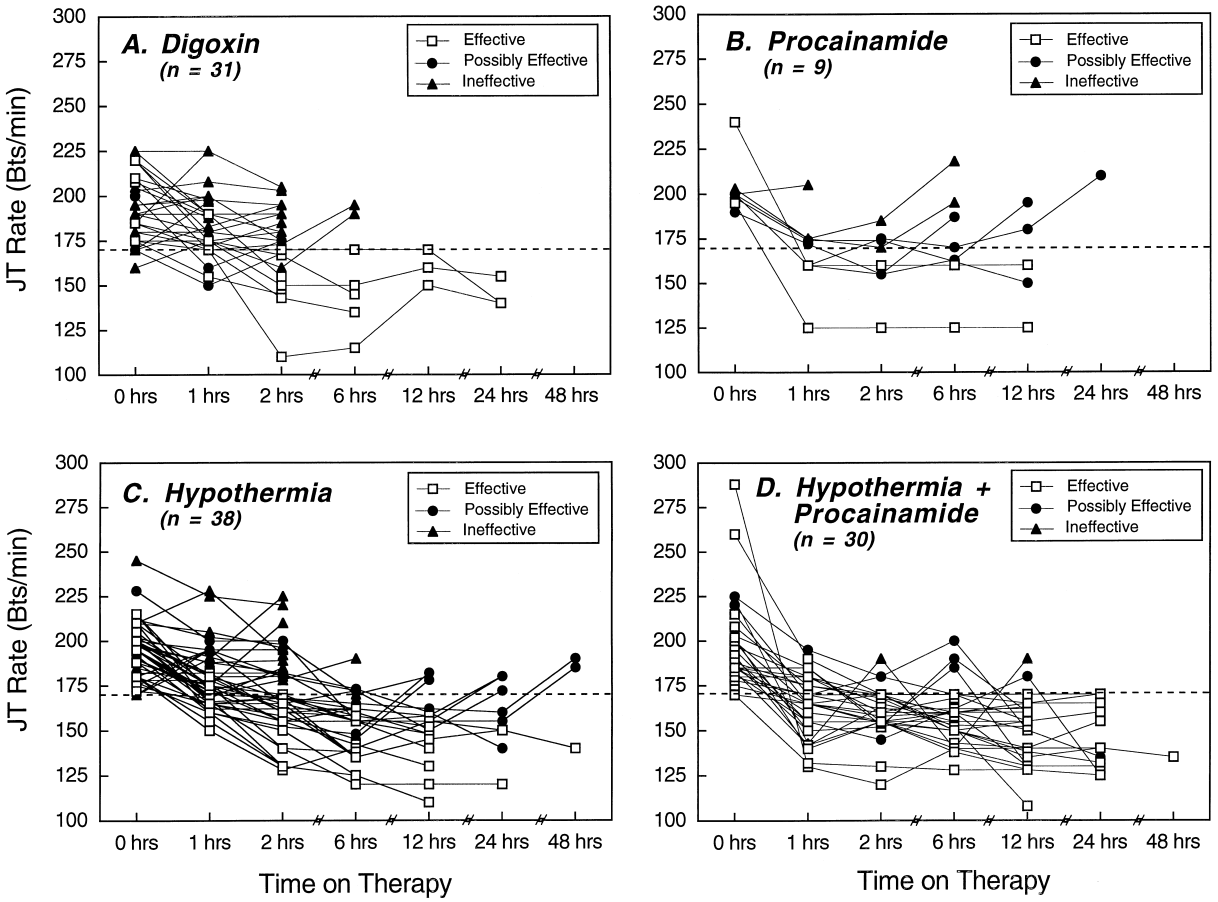


Figure 3. Time course for JT rates in individual patients during selected stages of therapy. The goal of treatment was to achieve a sustained reduction in the JT rate <170 beats/min **(dashed horizontal line)**. Patients remained on a given stage of therapy for variable periods and exited the stage at the time of either 1) resolution of JT; 2) escalation to another therapeutic stage; or 3) death. (See text for definitions of efficacy.)

IV agent were performed on 10 occasions early in the protocol and had little impact on JT. Procainamide was tested as an isolated therapy in nine patients. Despite the lack of long-term efficacy, there was a pattern suggesting acute JT slowing immediately after procainamide loading with a later rebound at 2 to 6 h, as shown in Figure 3B. The serum procainamide level for the "effective" or "possibly effective" subgroup was 3.9 ± 1.6 μg/ml, compared with 5.9 ± 1.6 μg/ml in the "ineffective" subgroup, making dose-related failure unlikely. Intravenous amiodarone was used in only one patient. A single loading dose of 5 mg/kg did not alter the JT rate, and long-term infusion was not tested.

Induced hypothermia was tested in 38 patients, with promising but inconsistent results (Fig. 2). A positive response was generally prompt (Fig. 3C), such that efficacy (or lack thereof) could be predicted within 1 or 2 h. Combined hypothermia and procainamide was tested in 30 patients and resulted in the most consistent JT control, with effective trials in 80% (Fig. 3D). The mean procainamide level for patients on combined therapy was 4.6 ± 2.5 μg/ml, which was nearly identical to that of patients receiving procainamide alone, despite the difference in efficacy.

The individual stages of therapy were evaluated statistically (Table 2) by comparison with the results for digoxin (assuming digoxin efficacy was negligible). Only normothermia and combined hypothermia and procainamide were significantly better than digoxin when only effective trials were considered as treatment success. If both effective and possibly effective trials were combined, normothermia, procainamide, hypothermia alone and combined hypothermia and procainamide were superior to digoxin.

Patient outcome. In 70 of 71 patients, JT was managed successfully. This was accomplished in 63 patients with an intervention scored as effective. In seven other patients who ultimately had good JT control, the therapeutic response was delayed beyond 2 h (n = 3) or was associated with later self-limited acceleration of JT (n = 4), and was thus considered possibly effective. The time to JT control for the total group was variable (range 1 to 69 h, mean 5.6 ± 11, median 3), in part reflecting the time spent early in our experience with trials of ineffective therapy. For the last 30 patients treated according to the refined protocol between 1990 and 1994, the time to control (mean 2.9 ± 2 h, median 2) was significantly shorter

Table 2. Efficacy of Therapeutic Stages for Rapid Postoperative Automatic Junctional Tachycardia

Therapy	Effective Trials	p Value*	Effective and Possibly Effective Trials	p Value*
Digoxin (n = 31)	7/31 (22%)	—	9/31 (29%)	—
⇓catecholamines (n = 16)	3/16 (19%)	NS	4/16 (25%)	NS
Normothermia (n = 17)	10/17 (59%)	0.01	13/17 (76%)	0.002
AV synchrony (n = 18)	5/18 (28%)	NS	10/18 (56%)	NS
Class IB, II, IV (n = 10)	0/10 (0%)	NS	2/10 (20%)	NS
Procainamide (n = 9)	2/9 (22%)	NS	5/9 (67%)	0.04
Hypothermia (n = 38)	12/38 (32%)	NS	22/38 (58%)	0.02
Procainamide and hypothermia (n = 30)	24/30 (80%)	< 0.001	29/30 (97%)	< 0.001

*Compared with results with digoxin.

(p = 0.04) than the time to control for the first 41 patients (mean 7.7 ± 15 h, median 3). The time to complete resolution of JT ranged from 2 to 168 h (mean 39 ± 38, median 36) and did not differ significantly between the original and refined protocol (mean 49 ± 42 h vs. 32 ± 34 h).

Treatment failed in one patient despite all levels of therapy. This patient was a 3-year old with a palliated hypoplastic left heart who underwent a Fontan procedure and had a postoperative course complicated by low cardiac output and metabolic acidosis. Despite hypothermia and procainamide, the patient ultimately died with continued low output and widely fluctuating JT rates of 140 to 190 beats/min. There were seven more hospital deaths that occurred in the setting of either resolved (n = 3) or well-controlled JT (n = 4), all of which had a cardiopulmonary explanation unrelated to rhythm status.

There were three complications potentially associated with therapy. Two children with effective JT control had acute ventricular fibrillation—one while on procainamide alone (serum level 3.0 μg/ml) and one while on combined therapy with procainamide (serum level 4.0 μg/ml) and hypothermia (33.8°C). Torsade de pointes was not documented in either case, and QT intervals were not prolonged. Both patients responded to electrical defibrillation and were maintained on the same JT therapy without recurrence of ventricular arrhythmias. One other patient, who was successfully managed with combined hypothermia and procainamide for 24 h, developed severe respiratory distress syndrome 48 h after return of normal sinus rhythm. The patient recovered but a contributory role for hypothermia in this atypical pulmonary process could not be ruled out.

Discussion

The central findings of this study are that 1) staged therapy (with an emphasis on combined hypothermia and procainamide in difficult cases) is an effective management approach for rapid postoperative JT; and 2) postoperative JT is strongly associated with young age, procedures that involve VSD closure and transient AV block. As newer treatment options such as intravenous amiodarone become more widely available, a reappraisal of JT management strategy has begun at many centers, including our own. The information from this report, which concentrated on "traditional" techniques for JT control, may serve the additional function of a comparison data base for prospective evaluation of alternate therapy.

Treatment of postoperative JT. No single stage of therapy was universally effective in this experience, and in fact, some commonly recommended JT treatments had minimal demonstrable value. Such was the case with digoxin, where the few effective trials probably represented the natural history of spontaneous JT resolution rather than drug effect. By eliminating digoxin and other nonproductive stages from the original protocol, a more efficient plan emerged, which significantly shortened the time to JT control. The observation that time to control could be shortened, without changing overall JT duration, strongly supports a true therapeutic effect for the refined protocol, rather than a natural history phenomenon.

There was practical merit to initiating treatment with simple measures, including reduction of exogenous catecholamines, pacing to restore AV synchrony and establishment of normothermia for febrile patients. Among these, only fever control seemed to have identifiable efficacy by statistical analysis. Nonetheless, these general measures could be tested quickly, had no obvious toxicity and were the only necessary therapy for 24% of our study group. All should be retained as first-line JT treatments, even if success is sporadic with catecholamine reduction and pacing. When AV synchrony was used in conjunction with drug therapy or hypothermia, or both, during this protocol, it was difficult to assess the isolated impact of pacing, but it seems likely that it was beneficial. This impression is reinforced by the study of Till and Rowland (6), who observed that AV synchrony provided additional hemodynamic benefit for patients already treated with drugs or hypothermia to reduce JT rates.

Hypothermia provided an effective reduction in JT rates for 21% of our patients, but ineffective trials remained common. Fortunately, failures of hypothermia could be predicted early into cooling, allowing therapy to be escalated without much delay. These data differ from the more optimistic results with hypothermia reported by Pfammatter et al. (14) in a series of six patients, perhaps because of the more rigorous definition of efficacy and the larger patient group used in the present study.

Combined hypothermia and procainamide provided the most reliable reduction of JT rate in our experience, and was ultimately the sole effective therapy for nearly half of the patients. It is notable that procainamide under normothermic conditions had less influence on the tachycardia, suggesting a temperature-dependent action (19) of this agent on the membrane channel(s) involved. Some investigators (10) have recently advocated use of high dose procainamide for JT, although review of drug levels in our patients did not support dose-related efficacy.

Successful JT slowing has also been reported with intravenous propafenone (8) and intravenous amiodarone. The single failure of amiodarone in our study would seem to be an unusual outcome compared with the experience of Raja et al. (11), who observed satisfactory reduction in JT rates after 2 h of therapy in 10 (62%) of 16 patients. Because of concerning alterations in hemodynamic variables and immune status that may accompany prolonged hypothermia (20,21), any agent that can function well under normothermic conditions needs to be seriously explored, assuming equivalent efficacy can be demonstrated. It would be hazardous to compare patient outcome in this report to published results for propafenone and amiodarone, because the definitions of successful therapy were not standardized. This issue should be addressed in a prospective manner.

In truly refractory cases of postoperative JT, ablation may have a role. Radiofrequency catheter ablation has proved useful in managing the congenital form of JT (22), and successful surgical cryoablation has been reported in one case of postoperative JT (15). Although it is sometimes possible to eliminate congenital JT without interrupting normal AV conduction (23), it is uncertain whether postoperative JT can be ablated without AV block. The risk of pacemaker dependency must be balanced against the self-limited nature of postoperative JT whenever ablation is considered.

Etiology of postoperative JT. Previous investigators have noted the tendency for JT to occur after surgery involving manipulation near the proximal conduction system, such as TOF repair and other forms of VSD closure (6,7,9,11,13,14). However, because these procedures account for a high percentage of cardiac operations in children, and because the number of patients with JT in previous reports was small, it has been difficult to test the statistical strength of this association. Analysis of the larger group in the current study strongly supports past clinical impressions and demonstrates that surgery incorporating VSD closure is significantly associated with JT. In contrast, JT was uncommon in the absence of VSD closure, regardless of whether the operation involved an intracardiac or extracardiac procedure. In addition, the high incidence of transient postoperative AV block noted in the JT group is consistent with the idea that trauma to the AV node or His bundle is key to the pathogenesis of JT, related either to suture placement during VSD closure or to stretch injury incurred while gaining surgical exposure. Finally, the association of younger age with JT may further support the concept of suture or stretch injury to the proximal AV conducting system, because there is less room for surgical manipulation in the smaller heart. Despite these findings, JT was also seen in 10 study patients (14%) in whom direct manipulation near conduction tissue seemed less likely: Fontan operation (n = 6), pulmonary artery banding (n = 2), Senning procedure (n = 1) and Norwood operation (n = 1). This inconsistency highlights the need for continued study of both the cellular mechanism and pathogenesis of JT. Potential directions for investigation include better quantitation of intraoperative trauma or stretch near the crux of the heart, as well as analysis of novel postoperative variables such as a recent report of elevated histamine levels among some patients with postoperative JT (24).

Study limitations. The major limitation of this study involves the self-resolving nature of postoperative JT, which confounds the distinction between natural history and therapeutic effect. In addition, therapy was tested according to a staged rather than a randomized design. We used consistent definitions of efficacy in an effort to minimize these issues, but recognize that the use of patients as their own controls during staged treatment limits the strength of the analysis.

An important question not addressed by this study is whether aggressive therapy is always indicated for rapid JT. Based on previous reports (7) and institutional experience suggesting a poor prognosis for untreated JT, we adopted a policy to treat all patients with rates >170 beats/min. This may or may not be a valid approach. Some centers prefer to follow patients with JT conservatively and have reported good outcomes with minimal intervention (25). Attempts to quantify the hemodynamic effects of JT and the benefits of therapy in the present study were difficult, but in agreement with most other investigators (5–14), we observed trends toward lower arterial blood pressure and higher atrial pressures when rapid JT began, which reversed when the rate was controlled and AV synchrony was restored. However, the more meaningful variables to measure in patients with JT involve mixed venous saturation or cardiac output, which were not part of the current or any previous JT study design. Cardiac output determination may be most critical when hypothermia is employed because increased systemic resistance is known to occur at temperatures of 31 to 35°C (20). We have now begun to rely on routine sampling of mixed venous saturations as a more objective measure of the necessity for (and effects of) JT treatment, rather than responding to rate alone.

Conclusions. Expeditious treatment of postoperative JT requires a large arsenal of treatment options. We describe one possible scheme that served a large group of patients well over the course of an 8-year trial, using a sequence of readily available drugs and techniques. Intravenous amiodarone may also be a reliable option, and data are now accumulating regarding its efficacy in this setting (11,26–28). Still lacking from all management plans is a therapy that restores sinus rhythm directly, rather than palliating the condition during the hours or days required until junctional automaticity resolves. Definitive therapy may remain elusive until the precise cause and cellular mechanism of JT are better understood.

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