Impact of junctional ectopic tachycardia on postoperative morbidity following repair of congenital heart defects


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Abstract

Objective: To determine the incidence of postoperative junctional ectopic tachycardia (JET), we reviewed 343 consecutive patients undergoing surgery between 1997 and 1999. The impact of this arrhythmia on inhospital morbidity and our protocol for treatment were assessed. Methods: We reviewed the postoperative course of patients undergoing surgery for ventricular septal defect (VSD; n = 161), tetralogy of Fallot (TOF; n = 114), atrioventricular septal defect (AVSD; n = 58) and common arterial trunk (n = 10). All patients with JET received treatment, in a stepwise manner, beginning with surface cooling, continuous intravenous amiodarone, and/or atrial pacing if the hemodynamics proved unstable. A linear regression model assessed the effect of these treatments upon hours of mechanical ventilation, and stay on the cardiac intensive care unit (CICU). Results: Overall mortality was 2.9% (n = 10), with three of these patients having JET and TOF. JET occurred in 37 patients (10.8%), most frequently after TOF repair (21.9%), followed by AVSD (10.3%), VSD (3.7%), and with no occurrence after repair of common arterial trunk. Mean ventilation time increased from 83 to 187 h amongst patients without and with JET patients (P < 0.0001). Accordingly, CICU stay increased from 107 to 210 h when JET occurred (P < 0.0001). Surface cooling was associated with a prolongation of ventilation and CICU stay, by 74 and 81 h, respectively (P < 0.02; P < 0.02). Amiodarone prolonged ventilation and CICU stay, respectively, by 274 and 275 h (P < 0.05; P < 0.06). Conclusions: Postoperative JET adds considerably to morbidity after congenital cardiac surgery, and is particularly frequent after TOF repair. Aggressive treatment with cooling and/or amiodarone is mandatory, but correlates with increased mechanical ventilation time and CICU stay. Better understanding of the mechanism underlying JET is required to achieve prevention, faster arrhythmic conversion, and reduction of associated inhospital morbidity. © 2002 Elsevier Science B.V. All rights reserved.

Keywords: Junctional ectopic tachycardia; Postoperative morbidity; Congenital heart defects

1. Background

Junctional ectopic tachycardia (JET) is a malignant arrhythmia of unknown aetiology, and a growing source of concern in the postoperative setting after repair of congenital heart defects (CHD). It has been reported after every type of surgical repair, but is most frequently observed after complete repair of tetralogy of Fallot (TOF) and surgery in the vicinity of the atrioventricular (av) node and the bundle of His. It frequently creates haemodynamic instability and requires aggressive management to allow survival. Despite a relatively high success rate in achieving a resolution of the arrhythmia with the current protocols for treatment, recovery is often prolonged, and the potential remains for death to occur. This study reviews the incidence of postoperative JET after surgery for selected congenital heart defects and its impact on inhospital morbidity. We present our current protocol for treatment and analyse its effect on stay in intensive care.

2. Methods

A retrospective analysis was performed on 343 consecutive patients undergoing surgery for certain congenital heart defects between January 1997 and December 1999 at the Cardiothoracic Unit of Great Ormond Street Hospital for Children NHS Trust, London, UK. Although JET has been...
reported after all types of surgical repairs, it is most commonly observed after surgical procedures which include closure of a ventricular septal defect, as well as after repair of tetralogy of Fallot. Accordingly, this study focused on those congenital heart anomalies requiring repair of a defect in the vicinity of the conduction system. The lesions targeted for analysis, therefore, included ventricular septal defect (VSD; \( n = 161 \)), tetralogy of Fallot (TOF; \( n = 114 \)), atrioventricular septal defect (AVSD; \( n = 58 \)), and common arterial trunk (\( n = 10 \)).

Surgery was performed by three surgeons using similar surgical technique, approaches to VSD closure, and relief of right ventricular outflow tract obstruction (RVOTO). Accordingly, cardiopulmonary bypass technique was standardized, including routine continuous arterial filtration, and modified ultrafiltration after discontinuation of bypass.

Data collection was based on patient charts, cardiac intensive care unit (CICU) records, and 12-lead surface or atrial electrocardiograms (ECG). Diagnostic ECG criteria for JET included: (1) a narrow QRS tachycardia (unless surgically induced right bundle branch block (RBBB) was evident) with a QRS rate between 170 and 230 beats per min; (2) AV dissociation with a ventricular rate faster than the atrial rate or retrograde ventriculo-atrial 1:1 conduction; and (3) unresponsiveness to adenosine, direct current (DC) cardioversion or overdrive pacing.

Upon diagnosis of JET, a stepwise treatment protocol was initiated until tachycardia converted to a stable rhythm, or else the patient died. Initial management commenced with avoidance of fever, followed by active surface cooling to facilitate active cooling, all patients were relaxed with packs placed around the head and trunk. Patients remained intubated and ventilated during treatment for the tachycardia. To avoid any stress-induced release of catecholamine, all patients were maximally sedated, using morphine (40 mg/kg per min) and midazolam (2–6 mg/kg per min). To facilitate active cooling, all patients were relaxed with vecuronium (2–4 mg/kg per min) infusions. Concomitant postoperative electrolyte imbalance was aggressively managed with appropriate infusions of potassium, calcium, and magnesium, to achieve levels above 4.0, 1.2, and 2.3 mmol/l, respectively. In the frequent cases of atrioventricular (AV) dissociation with haemodynamic instability, atrial or sequential pacing was initiated in an attempt to restore AV synchrony. This was achieved by using the temporary epicardial pacing wires placed at surgery. If the accelerated nodal rhythm further reduced cardiac output, with persistent metabolic acidosis, rate-control therapy with amiodarone was added. An initial intravenous loading dose of 25 mg/kg per min during 4 h was followed by a continuous infusion at 5–15 mg/kg per min. Monitoring of levels of amiodarone in the serum was performed when therapy was prolonged over several days.

Postoperative intensive care morbidity was considered in terms of hours of mechanical ventilation until successful extubation, and hours of total stay in the CICU until discharge to a paediatric cardiac stepdown unit. Transfer from the CICU to the stepdown unit may in some instances be dependent on staffing levels and bed availability, rather than patient acuity. For this reason, a Pearson’s correlations test between hours of ventilation and CICU stay was performed to assess the validity of this variable selection.

An analysis of variance was performed using hours of mechanical ventilation (VENT) and hours spent in the CICU as an end-point. Variables studied included the treatment modalities cooling (COOL), pacing (PACE), amiodarone (AMIOD), or a combination thereof. A one-way analysis of variance was performed to assess the effect of JET on hours of VENT and CICU with a two-sample t-test.

3. Results

Median age was 5.9 months (range 0.1–368.8 months). Per diagnosis, the youngest patients were those with common arterial trunk (median age = 0.4 months), followed by patients with AVSD (median age = 5.4 months), VSD (median age = 6.8 months), and TOF (median age = 11.2 months). Operative mortality was 3% (\( n = 10 \)), including four patients dying after repair of TOF, four after repair of common arterial trunk, and two after AVSD repair. In patients with JET, mortality was 8% (\( n = 3 \)), all of which occurred after TOF repair.

JET occurred in 37 instances (10.8%), most frequently after repair of TOF (\( n = 25 \); 21.9%), followed by AVSD (\( n = 6 \); 10.3%), VSD (\( n = 6 \); 3.7%), with no occurrences after repair of common arterial trunk. Per diagnosis, patients with common arterial trunk remained ventilated and in the CICU the longest (median = 220 and 270 h, respectively), followed by patients with AVSD (median = 90 and 118 h), TOF (median = 73 and 96 h), and VSD (median = 17 and 37 h).

While standard criteria for extubation are universally accepted, the indications for transfer from the intensive care to a stepdown unit are less so. Despite this potential variability, the Pearson’s correlations between hours of VENT and CICU stay was 0.988 (\( P < 0.0001 \)). The diagnosis of JET, and its associated strategies for treatment, significantly increased the mean duration of postoperative mechanical ventilation as compared to controls without JET (187 ± 25 versus 83 ± 12 h, respectively, \( P < 0.0001 \)). Correspondingly, the mean duration of CICU stay was prolonged in patients with JET, as compared to controls without (210 ± 25 versus 107 ± 13 h, respectively, \( P < 0.0001 \)) (Table 1).

Cooling was performed in 47 patients, including 34 patients with TOF (29.8%), one with common arterial trunk (10%), five with VSD (3.1%), and seven with AVSD (12.1%). Atrial or sequential pacing was performed in 21 patients, including 16 patients with TOF (14%), three with VSD (1.9%) and two with AVSD (3.4%), but in none
after repair of common arterial trunk. Amiodarone was initiated in 22 patients, including 15 patients with TOF repair (13.2%), five with VSD (3.1%), and two with AVSD (3.4%). Fourteen patients required all three modalities, including nine with TOF, three with VSD, and two with AVSD. Ten children needed cooling and pacing (five with TOF and five with AVSD). Seven children needed cooling and amiodarone (five TOF + two VSD). Cooling alone resulted in a lower heart rate until spontaneous arrhythmic conversion in 21 patients, and two patients benefited from atrial pacing alone.

According to our protocol, cooling and antiarrhythmic treatment with amiodarone were discontinued 24–48 h after conversion to sinus rhythm with a heart rate in normal limits. This was achieved in 34/37 patients, giving a rate of success of 91.9%. The rate of mortality in patients with JET limits. This was achieved in 34/37 patients, giving a rate of mortality in patients with TOF, giving a JET-mortality, or protocol failure rate, of 12% in patients with TOF.

Using a linear model with analysis of variance, cooling was initiated at the slightest suspicion of JET. Thus, 47 patients underwent cooling, of whom only 37 were subsequently established to have JET. Similarly, five patients received amiodarone, and three patients were atrially paced, without surface or atrial-lead ECG confirmation of JET. This somewhat aggressive early initiation of management may be clarified in light of the finding that 21/37 (56.8%) of patients with a confirmed diagnosis of JET responded to cooling alone, without further escalation in the protocol for treatment.

Using a linear model with analysis of variance, cooling was significantly associated with a prolongation of VENT time by 74 ± 30 h (P < 0.02) and CICU stay by 81 ± 32 h (P < 0.02). Treatment with amiodarone was significantly associated with an increase of mechanical ventilation by 274 ± 133 h (P < 0.05) and length of CICU stay by 275 ± 142 h (P < 0.06) (Table 2).

4. Discussion

Junctional ectopic tachycardia is a potentially life-threatening, although eventually self-limiting, arrhythmia. It occurs rarely in a spontaneous congenital form, but most commonly arises in the postoperative setting of surgery for congenital heart diseases [1–14]. Villain et al noted a familial variant in half of their patients with congenital JET [4], and mortality in these patients has been as high as 35% despite treatment.

The true incidence of postoperative JET is probably underreported, and is estimated to be between 1 and 22% after repair of CHD [7,14]. The precise aetiology of JET is unknown, but it is believed to be the result of enhanced automaticity in the bundle of His, either in its right atrial or right ventricular portion, promoted by haemorrhage into the conduction tissues [1]. Successful radiofrequency ablation is sometimes difficult in refractory JET, leading some authors to suggest a ‘left-sided’ bundle of His, or a JET focus emerging from the left side of the ventricular crest, before propagating to the His bundle [4,9]. Autopsy studies of surgical specimens with JET have disclosed streaks of haemorrhage penetrating the atrioventricular bundle and node on the left side of the ventricular crest, in addition to direct suture damage within the central fibrous body [1,12]. It is postulated that disruption of conduction tissue, either by direct trauma or penetrating blood and interstitial inflammatory cells, may result in an irritable focus leading to JET [1].

The incidence of postoperative JET after repair of TOF is relatively high (21.9%) in our series. Speculating on causative surgical, anatomical or cardiopulmonary bypass factors in the genesis of JET, a critical analysis of the complete patient population (n = 343) was performed [15]. Specifically, variables looking at surgical techniques of VSD closure, and approaches towards relief of RVOTO were studied. After univariate analysis, followed by multivariate analysis including 16 perioperative and operative variables of surgical technique, approaches to repair, and cardiopulmonary bypass data, only resection of RVOTO muscle bundles (versus sharp division), higher temperatures on cardiopulmonary bypass, and transatrial relief of RVOTO, were significant and independent risk factors for JET (P < 0.0001, P < 0.03, and P < 0.05, respectively) [15].

The diagnosis of JET is best established by a 12-lead surface ECG at 50 mm per s, followed by an atrial-lead ECG, performed during the tachycardia [3–8]. The temporary epicardial atrial pacing wires routinely placed during surgery make an atrial ECG readily available. It is important to exclude a sinus or supraventricular tachycardia by an adenosine challenge, and atrial pacing to assess atrial capture [3]. JET is characterised by a narrow QRS complex (unless surgical right bundle branch block has occurred), with atrioventricular dissociation, most often creating haemodynamic instability from loss of atrial contraction and its contribution to cardiac output [1,3,5,7,8]. The arterial and venous pressure wave forms will correspondingly

Table 1

<table>
<thead>
<tr>
<th>Effect of JET on inhospital morbidity^a</th>
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<tr>
<td>Vent (h)</td>
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<tr>
<td>Jet (n = 37)</td>
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<td>Controls (n = 306)</td>
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<td>P &lt; 0.0001</td>
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^a Vent, ventilation; h, hours; and CICU, cardiac intensive care unit.

Table 2

<p>| Effect of treatment protocol on inhospital morbidity^a |
|-------------|--------|</p>
<table>
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<tr>
<th>Vent (h)</th>
<th>CICU (h)</th>
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<tr>
<td>Cooling</td>
<td>74 ± 30 (P &lt; 0.02)</td>
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<tr>
<td>Amiodarone</td>
<td>274 ± 133 (P &lt; 0.05)</td>
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^a Vent, ventilation; h, hours; and CICU, cardiac intensive care unit.
In JET, before and after cooling.

![Diagram](https://academic.oup.com/ejcts/article-abstract/21/2/255/2755203)

**Fig. 1.** Beat-to-beat variability of arterial and venous pressure wave forms in JET, before and after cooling.

Demonstrate a beat-to-beat variability (Fig. 1). In the great majority of cases (90%), the atrial rate is slower than the ventricular rate, but P waves may be retrograde with ventriculo-atrial 1:1 conduction in 10% of cases [1,7]. In common with other automatic tachycardias, JET typically presents in a ‘warm-up’ and ‘warm-down’ pattern, with progressive acceleration into, and deceleration out of the tachycardia [3,6–8]. The QRS rate is above the 98th percentile for the age of the patient, typically between 170 and 230 beats per min. JET is unresponsive to overdrive pacing or direct current (DC) cardioversion [3,6–8]. Once the rate of the tachycardia is slowed by cooling or medication, atrial pacing becomes a useful adjunct to support cardiac output. Atrial pacing conducts normally to the ventricle, thus yielding a normal QRS morphology and AV synchrony [3,6,8].

Aggressive management is required in the postoperative setting, as the arrhythmia frequently induces haemodynamic instability through atrioventricular dissociation. It typically feeds on itself in a vicious circle pattern, as the lack of AV synchrony and the high ectopic rate lead to diminished cardiac output, leading to a reflex increase in adrenergic tone, which further accelerates the arrhythmia.

Concomitant postoperative electrolytic imbalance is frequent, and requires appropriate treatment [14,16]. Hypokalemia, hypocalcemia and hypomagnesemia must be managed to keep levels of potassium above 3.5–4.0 mmol/l [13,14], levels of calcium above 0.8–1.2 mmol/l [13,14], and levels of magnesium from 1.5 to 2.3 mg/dl [13,14]. Recent evidence suggests that magnesium depletion in the postoperative setting may significantly increase the risk of developing JET [13]. Dorman et al. presented data from 28 patients undergoing surgical repair for CHD in which magnesium supplementation was prophylactically given in a double-blinded fashion, directly after discontinuation of CPB. JET did not occur in those receiving magnesium, as compared to an incidence of 27% in patients given saline, before the study was discontinued for ethical reasons [13]. Prophylactic supplementation with magnesium, therefore, may have a role in preventing JET, and is performed in our unit. Acidosis and hypovolemia are commonly associated and should be addressed. Endogenous and exogenous catecholamines may worsen the tachycardia. Accordingly, cardiac inotropic support should be reduced, and even avoided, if possible [3]. Meticulous medical and nursing care, including adequate sedation and avoidance of vagolytic therapies, should reduce patient stress and any risk of endogenous catecholamines [3]. Hyperthermia must be avoided [7], and active surface cooling to 32–35°C is a first-line integral part of treatment during JET [3,5,8]. Aggressive antiarrhythmic intervention usually achieves stabilisation, and subsequent spontaneous termination is the rule, commonly occurring between 2 and 8 days [3,5].

Many antiarrhythmic therapies have been employed with variable rates of success. Beta blockade with propranolol may provide sufficient adrenergic block, but is limited by its negative inotropic effect, which is undesirable in a postoperative setting [3]. The class IC agents flecainide, encaïnide, and propafenone, have been successfully used in management [3,8,17]. Many centers, including our own, favour the class III agent amiodarone as the agent of choice [6,7,18]. Favourable pharmacokinetics allow various regimens for administration, including intermittent bolus doses or loading, plus continuous infusion protocols. Upon achieving sinus rhythm, amiodarone is discontinued after a minimum of 24 h of drug therapy, but the patient remains continuously monitored for a further 24 h. As this is a self-limiting disorder which usually abates in 2–8 days, long-term therapy with amiodarone is unwarranted if no further recurrence occurs during inhospital stay [7].

Intractable JET refractory to all antiarrhythmic medication or pacing methods may be aborted by transcatheter ablation of the His bundle [3,9,17]. Although radiofrequency ablation has been successfully employed in infants and children with the congenital form of JET [19,20], its use in self-limiting postoperative JET should be restricted to refractory cases, as there may be an unacceptably high risk of complete heart block [17].

Extracorporeal membrane oxygenation (ECMO) as an emergency procedure has been successfully used in patients with JET refractory to all conventional treatments outlined earlier [10]. The self-limited nature of JET makes ECMO support a reasonable adjunct to conventional therapy that allows circulatory assistance, and precise control of hypothermia, until spontaneous recovery from the arrhythmia occurs. The various risks and contraindications pertain-
ing to bleeding, exposure to additional blood products, neurological damage, and infections, must be outweighed by the life-saving nature of ECMO in this setting.

In conclusion, JET is an increasingly recognised malignant arrhythmia arising in the postoperative setting after surgery for congenital heart disease. Multiple protocols exist for treatment, and most have demonstrated their efficiency in sustaining patients until spontaneous arrhythmic conversion. Nearly all protocols call for profound sedation, paralysis, maintenance of mechanical ventilation, avoidance of fever, and surface cooling in the first instance. We found this initial step to be therapeutic in itself in a majority of cases, without need for further escalation to pacing or anti-arrhythmic therapy. In five patients, amiodarone may have been required anyway to treat a refractory supraventricular arrhythmia, despite overdiagnosis of JET. Accordingly, temporary pacing in three patients without a confirmed diagnosis of JET achieved atrioventricular synchrony and improved cardiac output. Nonetheless, the distinction between true JET and other atrial arrhythmias is essential, as excessive therapy may result in unnecessarily prolonged ventilation time and CICU stay, both of which carry their own potential morbidity, mortality, and cost considerations. Strict interdisciplinary collaboration between surgeons, cardiologists, and intensive care physicians is necessary to assess this difficult arrhythmia, in terms of both diagnosis and potential escalation in management. A better understanding of the underlying mechanisms of JET after surgery for congenital heart disease may lead to a reduction or avoidance of this malignant arrhythmia, and to faster and more efficient conversion in the remaining cases.

5. Study limitations

Although more frequently encountered after cardiac surgery involving closure of a VSD, JET may occur after the repair of virtually every congenital heart defect, even extracardiac ones [17]. However, Walsh and colleagues [17] reported a negative association of JET after surgery to close defects within the oval fossa, after coarctation or aortic arch repair, pacemaker implantation, and confection of systemic-to-pulmonary arterial shunts. Accordingly, in this study, patients with simpler intracardiac or extracardiac lesions were not included. Also, it may be argued that many complex congenital defects include closure of a VSD amongst a long list of other anomalies to correct. However, these were not analysed in the current study, in an attempt to minimize the potential effect of a wide spectrum of morphology.

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