Diagnosis and management of junctional ectopic tachycardia in children

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ABSTRACT

Junctional ectopic tachycardia (JET) is more common in its postoperative form. A thorough understanding of its etiology, pathophysiology, and management strategies is essential. Classically, postoperative JET is considered to arise from surgical trauma. Genetic susceptibility and an intrinsic morphologic/functional defect in the conduction system inherent in congenital heart diseases likely play a significant role. The devastating effects on postoperative hemodynamics warrant prompt attention. A multipronged management approach with general measures, pharmacotherapy, and pacing has decreased morbidity and mortality. Amiodarone and procainamide remain the preferred drugs, while ivabradine appears promising. Carefully planned randomized trials can go a long way in developing a systematic management protocol for postoperative JET.

Keywords: Congenital junctional ectopic tachycardia, junctional ectopic tachycardia, pediatric cardiopulmonary bypass, postoperative arrhythmia, postoperative complications

INTRODUCTION

Junctional rhythms originate from the atrioventricular (AV) junction. They encompass a wide spectrum of arrhythmias with variable clinical presentations, significance, and treatment strategies. Junctional ectopic tachycardia (JET) was first described by Coumel *et al.* in 1976 as a distinct entity and is an automatic focus rhythm most commonly seen in children.[1]

JET is recognized on the surface electrocardiogram (ECG) by a narrow QRS tachycardia, with AV dissociation (although AV association may be rarely present) and a faster ventricular rate.[2] JET occurs in two distinct clinical settings: (i) a permanent form, a primary idiopathic disorder during infancy called the congenital JET, and (ii) a transient form, in children and also in adults immediately after surgery for congenital heart disease (CHD), called the postoperative form.[3-5] Isolated cases of JET following viral myocarditis have also been reported.[6,7]

CONGENITAL JUNCTIONAL ECTOPIC TACHYCARDIA

The congenital form of this pathology is discussed briefly however is not the main focus of the review. Congenital JET is rare, accounting for <1% of pediatric arrhythmias, and usually occurs in infants <6 months of age. It has a familial predisposition in around 50% of cases, commonly presents with cardiomegaly, congestive heart failure, and echocardiographic

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evidence of left ventricular dysfunction, and has a mortality rate as high as 34%.[8,9] A pharmacologic approach is regarded as the first-line therapeutic option for infants with symptoms, reduced ventricular function, or rapid heart rate.[9] Digoxin, amiodarone, propranolol, and flecainide have been used alone or in combination.[9-12] More recently, ivabradine, the selective and specific inhibitor of the cardiac pacemaker current (If), has been found useful in resistant cases.[12,13] Successful transcatheter ablation has been reported with preservation of normal AV conduction and is suggested for drug refractory cases.[10,14-19] A few cases of sudden death have been attributed to evolution to complete heart block, unrelated to ablation procedures.[8,20] Prophylactic pacemaker implantation is controversial.[5] Permanent pacemaker implantation is largely indicated for AV block secondary to ablation procedures.[11] Documented AV block, either on ECG, Holter or on transesophageal electrophysiological test, might warrant permanent pacemaker implantation. Improved outcomes with a mortality rate of 4% have been reported with current therapies, which mainly consist of medical management (primarily amiodarone), radiofrequency and cryoablation, and permanent pacemaker implantation.[11]

POSTOPERATIVE JUNCTIONAL ECTOPIC TACHYCARDIA

This is the most common form of JET, usually occurring in the first 24–48 h after surgery.[21] Postoperative JET is associated with increased risk of morbidity and mortality. Grosse-Wortmann *et al.* described JET as typically occurring 2–8 days after cardiopulmonary bypass (CPB).[21] JET has also been rarely documented after the 1st week of surgery.[22] The mean cardiac intensive care unit stay and the mean mechanical ventilation time have been reported to be high for patients who developed JET in the postoperative period compared to those who did not.[23] JET is a self-limiting disorder that usually resolves within 1 week. Onset is at a time when the systolic and diastolic function of the heart is generally impaired. The rapid heart rate and loss of AV synchrony lead to decreased cardiac output and increased central venous pressure, which result in multiorgan dysfunction. The reduced cardiac output stimulates the adrenergic system, which in turn further accelerates the heart rate. This rapidly evolves into a vicious hemodynamic spiral.[24,25] Hence, postoperative JET warrants prompt treatment.[24] Incidence, mostly based on surface ECGs, varies from 1% to 10%.[21,22,26-31] Mortality was high early on near 40%.[32,33] Later reports showed significant improvement in prognosis. A Spanish group reported a mortality of 8.3%.[34] No deaths were reported to be related to JET in other later studies, likely from prompt identification and multipronged management approach.[22,28,35,36] The etiopathogenesis, diagnosis, and management are reviewed below.

ETIOPATHOGENESIS

Abnormal automaticity is considered to be the primary underlying mechanism.[37] Much research has focused on the risk factors for developing postoperative JET, including type of underlying heart disease, surgical procedures and CPB times, genetics, and electrolyte disturbances.

Conflicting literature exists as to the nature of surgeries associated with the highest incidence [Table 1], supporting the multifactorial model of etiology.[21,22,27,29] Although the incidence is higher in some diseases than others, the nature of surgeries associated with higher incidence varies from center to center. This can partly be explained by the type/spectrum of surgeries each center is performing. Even for the same lesion, the incidence varies from center to center.[26,30]

Factors contributing include direct surgical injuries such as myocardial incision, sutures close to the conduction system, trauma to conduction system during surgical handling, and cardiac volume and pressure fluctuations.[29,30,39,40] CPB with ischemia–reperfusion and the related cellular biochemical effects, electrolyte shifts, and catecholamine administration can affect the stability of the cellular membrane and result in an increased myocardial irritability and automaticity.[41] Inflammatory mediators released in the early postoperative period can alter the membrane potential of myocytes.[42] This can also facilitate micro-reentry within the atrium and the AV node.[21] The effects of CPB on release of inflammatory mediators such as histamine and eosinophil cationic protein by basophils and their relation to postoperative arrhythmias including JET were shown by Seghaye *et al.*[43] Estradiol inhibits the release of inflammatory cytokines. It is supposed to have a protective effect against arrhythmias.[21,44] Younger age is one of the commonly implicated risk factors for postoperative arrhythmias.[21-23,40,45] Smaller hearts are subjected to more trauma during surgery, direct and indirect. The higher complexity of corrective infant heart surgeries could also explain higher incidence (14.3%) after infant heart surgery.[46] Several studies have also shown that more than the anatomic substrate or the type of surgery, longer CPB times are the predictive of postoperative arrhythmias including JET.[26-28,39] Complexity indirectly indicates longer ischemic times, more reperfusion injury, and more inflammatory response "Surgical procedures remote from the AV node like resection of sub aortic stenosis , atrial septal defect closure and cardiac transplantation has also been associated with

Table 1: Surgeries reported to be associated with high incidence of junctional ectopic tachycardia

Study	Surgeries most commonly reported to be associated with JET
Batra *et al.*[27]	Arterial switch operation 23% (*n*=13), AV canal repair 21% (*n*=19), and Norwood repair 20% (*n*=10)
Delaney *et al.*[28]	Complete AV septal defect repair 72% (*n*=11), arterial switch 62.5% (*n*=8)
Pfammatter *et al.*[38]	Ventricular septal defect 30% (*n*=75), tetralogy of Fallot 35% (*n*=52), AV canal 47% (*n*=45)

AV: Atrioventricular, JET: Junctional ectopic tachycardia

JET."[21,22] Further, extra cardiac procedures procedures without CPB such as extracardiac Fontan , hemi-Fontan and Blalock-Taussig shunt have also been associated.

Genetic susceptibility and an intrinsic morphologic/functional defect in the conduction system inherent in a specific CHD unrelated to the iatrogenic factors, but predisposing to JET, are plausible.[22,29,45,47] Angiotensin-converting enzyme deletion polymorphism is reportedly associated with more than two times increase in the odds of developing JET, suggesting the potential role of renin–angiotensin–aldosterone system.[37,45]

Hyperpolarization- activated cyclic nucleotide–gated (HCN) channel upregulation may play a role in the genesis of JET. They are found not only in the SA node AV node but also in the conduction system of the myocardium which includes His bundle and Purkinje fibers. Although their presence is of no physiological significance in the normal myocardium, HCN channels are upregulated in remodeling of ischemia, hypertrophy, and heart failure.[48] In rat studies, HCN channels upregulated within a day after myocardial infarction.[49] A similar change is plausible after CPB. Notwithstanding CPB, CHD patients have hemodynamically significant lesions, which negatively affect the ventricle by remodeling, and can upregulate HCN channels in the conduction system, predisposing them to arrhythmias. Triggers and modulators such as electrolyte imbalance and excess catecholamine acting on the electrically remodeled ventricle with upregulated HCN channels might explain JET after surgeries without any possibility of mechanical injury to conduction system and no CPB, such as Fontan and the Blalock-Taussig-Thomas shunt.

Electrolyte imbalances, prominently hypomagnesemia, have been implicated. The literature on hypomagnesemia and JET is conflicting.[22,28,38,39,44,50-53] Nevertheless, the role of magnesium in preventing JET was reinforced by a prospective randomized double-blind study of the effect of magnesium supplementation on arrhythmias after CPB. This study was terminated prematurely because of the unacceptable incidence of hemodynamically significant JET in the placebo group.[54]

DIAGNOSIS

JET can be diagnosed on surface ECG. "JET is best defined as a tachycardia originating from the AV junction with gradual onset, gradual offset, rate variability, narrow QRS

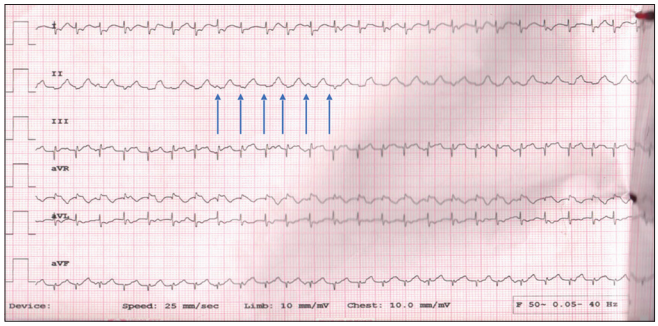

Figure 1: Postoperative junctional ectopic tachycardia. Limb leads only. Ventricular rate of 190/min. P waves, indicated by the blue arrows might not be readily identified on the surface electrocardiogram

complex, usually with a ventricular rate faster than the atrial rate and AV dissociation."[11,38] The heart rate in JET is variably defined as a junctional rate >95th[11,55,56] or 98th[29,57] percentile of heart rate for age or more than 170 beats per minute.[11,29,50] The latter definition is handy from a practical viewpoint. Invasive electrophysiological study is rarely needed. The electrophysiological hallmark of JET is that each QRS complex is preceded by a His bundle depolarization. Except when there is underlying conduction system disease, the His ventricular interval is normal. Ventriculoatrial conduction block and AV dissociation are the common findings, but ventriculoatrial conduction can sometimes be seen. JET has spontaneous initiation and termination. As an automatic focus tachycardia, JET cannot be terminated by direct current cardioversion, by critically timed premature beats, or by overdrive pacing. Failure to convert the "SVT" with DC cardioversion should be taken as evidence in favor of JET or other automatic tachycardias (such as ectopic atrial tachycardia). Temporary atrial wires can be used to display an atrial lead ECG for additional information[29] [Figure 2 - atrial electrogram]. This is helpful when atrial depolarization is not visible on surface ECG, the relation between atrial and ventricular electrical activity is unclear on surface ECG, wide QRS tachycardia needs to be better defined, or whenever the mechanism of arrhythmia is unclear.[58] Adenosine will not terminate the arrhythmia but block conduction to the atria, resulting in VA dissociation. In patients with clear presence of AV dissociation during the arrhythmia, there is little diagnostic benefit to adenosine. However, in those with 1:1 conduction to the atrium, adenosine can be used to unmask AV dissociation and confirm the diagnosis of JET.

The ventricular rate can be irregular with sinus capture beats when appropriately timed *P* waves conduct to the ventricles. Rarely, there can be 1: 1 retrograde VA conduction, with a ventricular rate equal to atrial rate, and when this happens, it may be difficult to distinguish JET from other forms of supraventricular tachycardia. If P waves are not identifiable, ECG can mimic appearance of atrial fibrillation. The QRS complex is similar in morphology to baseline sinus beats and can be wide when there is underlying bundle branch block on baseline ECG. If so, it may be difficult to distinguish JET from ventricular tachycardia.

MANAGEMENT

A recent European survey reflected the widely practiced multipronged approach. The antiarrhythmic of choice was amiodarone, used in varying sequential order with therapeutic hypothermia and pacing.[59] Table 2 summarizes published management strategies from the literature. So far, no significant differences in hard outcomes between the small groups have been detected. Given the widespread acute effects and probable long-term sequelae of any period of low cardiac output, it seems prudent to start early aggressive therapy to control/convert JET before signs of low cardiac output ensue. Patients with low cardiac output at onset of JET were found to have a predilection for amiodarone unresponsiveness. These patients as well as patients deemed to be otherwise at high risk of same would require combined therapy with hypothermia and pharmacological agents [Figure 3]. Restoration to sinus rhythm is the ideal target of treatment. This might not always be readily achievable. A decrease in heart rate adequate for hemodynamic improvement or one that allows atrial or AV sequential pacing, and thereby improving cardiac output is generally considered as success of treatment . This is in the background that JET is a self limiting disorder, resolving in 48-72 hours or a week.[24,35,60] Inadequate response warrants consideration of newer agents such as ivabradine/sotalol. Stable patients can be started on either hypothermia or pharmacotherapy but need to be vigorously monitored for response and treatment escalated as needed. It was noted in a small group of patients that when the heart rate exceeded 200 beats/min for 5 h, congestive heart failure and clinical signs of low cardiac output manifested.[32] The window from onset of JET to low cardiac output syndrome is highly variable. Hence, it is inappropriate to wait for more than a few hours for response to initial treatment modality. General measures, pacing, hypothermia, and pharmacotherapy including newer agents ivabradine/sotalol are discussed in more detail below.

General measures

General measures to be instituted include correction of fever, electrolyte abnormalities, with specific emphasis on magnesium, anemia and hypovolemia, optimizing sedation and pain control, and reducing inotropic drugs to the minimum tolerated dose.[22,28,35]

Although catecholamines can be considered as a group and tachycardia as a class effect, a lower threshold for

Table 2: Management strategies mentioned in literature

Author	Management protocol	Comments	Mortality
Hoffman *et al.*[22]	Staged approach - general measures first (avoidance of hyperthermia -antipyretic agents and removal of external sources of radiant warming, sedation, and minimizing catecholamines) and specific therapy (amiodarone/pacing/active cooling) upon physiologic signs of low cardiac output	13/33 (39.4%) of the patients received specific therapy	No death attributable to JET
Batra *et al.*[27]	Conventional measures (cooling and sedation) as first-line intravenous amiodarone if no response	48% (*n*=13) responded to conventional measures; 48% (*n*=13) required intravenous amiodarone therapy	1 death attributable to JET
Delaney *et al.*[28]	General measures and atrial overdrive pacing through temporary atrial wires to restore synchrony if the ventricular rate permitted. Amiodarone infusion if ventricular rate was too rapid for effective overdrive pacing	Study was aimed mainly to determine incidence and risk factors	No death attributable to JET
Walsh *et al.*[50]	Staged approach involving A reduction of catecholamines Correction of fever Atrial pacing to restore synchrony Digoxin Phenytoin or propranolol or verapamil Procainamide or hypothermia Combined procainamide and hypothermia	Last 30 patients achieved rapid control with early hypothermia and procain amide in addition to general measures and AV synchrony Treatment effective in 70/71 patients. General measures alone effective in 24%, hypothermia effective in 21%, Combined hypothermia and procainamide provided most reliable reduction of JET rate was the sole effective therapy for nearly 50%	1 death attributable to JET
Kovacikova *et al.*[35]	Amiodarone as first line after general measures if they failed to bring down the rate below 170 beats/min, hypothermia as second line	Amiodarone was effective in 18/40 patients (45%), 22/40 patients (55%) required escalation therapy to hypothermia	No death attributable to JET

AV: Atrioventricular, JET: Junctional ectopic tachycardia

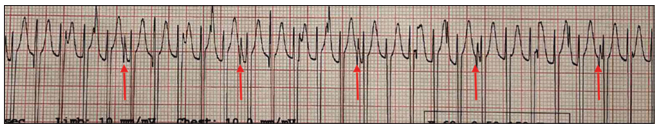

Figure 2: Atrial electrogram from a patient with junctional ectopic tachycardia, demonstrating atrioventricular dissociation. P waves on the atrial electrogram are marked with red arrows

decreasing dose or discontinuing/substituting dopamine can be adapted keeping in mind its proven association in a multivariate analysis.[22] The same study found an association with milrinone in univariate analysis. Milrinone and other vasodilators may accelerate rate by production of endogenous catecholamines. The association could also be attributed to complex nature of surgeries, where milrinone use is frequent.[22]

It is important to avoid DC cardioversion if possible. It does not "convert" JET to normal rhythm and causes unnecessary cardiac trauma in a vulnerable postoperative child.

The importance of atrioventricular synchrony and its restoration by pacing

Most treatment methods described are better at slowing the JET rate rather than converting it to normal sinus rhythm. Even if rate is reduced, AV dissociation or ventriculoatrial conduction remains, resulting in atrial contraction almost coincident with ventricular contraction. Cardiac output can be significantly increased in the sick heart by the addition of atrial contraction at correct time in cardiac cycle even though cardiac output in small children is predominantly rate dependent.[61] As discussed previously, slowing of JET to an acceptable heart rate that allows hemodynamic improvement with atrial or AV sequential pacing is often defined as success of any particular form of therapy for JET. This also highlights the fact that restoring AV synchrony by using different modes of pacing is an integral part of management of JET.[25,34-36,61-63] It has been demonstrated that several techniques of individually optimized temporary dual chamber pacing, leading to optimal AV synchrony and/or synchronous ventricular contraction, can be successfully used to improve hemodynamics in patients with heart failure and selected dysrhythmias after congenital heart surgery.[64] Atrial pacing, AV sequential pacing, R wave synchronized atrial pacing, and paired ventricular pacing are all modalities that can be helpful in achieving hemodynamic stability.

Atrial pacing at a rate higher than JET rate can restore AV synchrony in a subset of patients with JET, accepting the disadvantages of a higher rate. In most patients, atrial pacing is successfully conducted to the ventricles through the AV node restoring AV synchrony. In rare cases, impaired AV conduction may be present and is revealed after institution of atrial pacing making AV

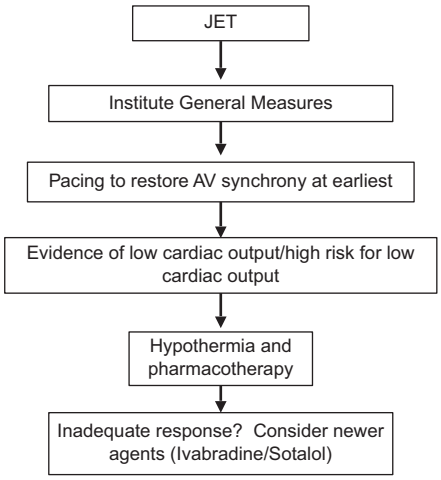

Figure 3: Approach to management of postoperative junctional ectopic tachycardia

resynchronization by atrial pacing impossible. In such cases, AV sequential pacing should be used. Since, by necessity, the paced rate will have to be higher than the JET rate, pacing may be disadvantageous unless hypothermia/pharmacological measures bring down the JET rate. Other drawbacks while using AV sequential pacing include ventricular dyssynchrony and contractile incoordination induced by ventricular pacing. R wave synchronized atrial pacing was suggested to overcome the disadvantages of AV sequential pacing. This was first reported to be achieved by using two stimulators in concert – one to receive input from ventricle and other to pace the atrium after a preset delay.[61] Later, in the same study, a dual chamber analyzer (Medtronic) was used, the ventricular temporary pacing wires were connected to the atrial input, and the atrial wires were attached to the ventricular output in the VDD pacing mode with an AV delay set so that the atrial pacing stimulus preceded the subsequent QRS complex by an appropriate interval.[61]

Janousek *et al.* successfully used a modified, commercially available temporary dual chamber pacemaker for R wave synchronized atrial pacing. R wave synchronized atrial pacing was performed using the VAT mode with inverse connection of the pacing wires (effectively AVT mode), short postventricular atrial refractory period (100 ms), and long AV (effectively VA) delay. Complications, especially pacemaker-mediated tachycardia, were easily stopped by prolongation of AV (effectively VA) delay. Atrial flutter induced by asynchronous atrial pacing during the VAT (effectively AVT) mode was managed by overdrive pacing. Significant hemodynamic benefit was achieved due to optimal AV resynchronization at intrinsic heart rate and spontaneous ventricular activation sequence.[65]

Paired ventricular pacing is another modality that can be used to lower effective ventricular rate in patients

with postoperative JET.[66-68] Pairs of stimuli are applied, second one shortly after the effective refractory period of first one. Electrical activation occurs from the second stimulus, but no mechanical contraction. This produces a postextrasystolic potentiation type effect on the next first stimulus, increasing cardiac output. Pacing protocols are generated using an electrophysiology simulator, which would be connected to ventricular leads. Disadvantages include the risk of ventricular fibrillation, inability to re-establish AV synchrony, and induction of ventricular dyssynchrony by single site ventricular pacing. Practicality is limited as it would require a sophisticated EP lab style simulator. Paired ventricular pacing can be reserved for acute management of patients with a high junctional rate precluding R wave synchronized atrial pacing or AV sequential pacing.[65]

Hypothermia

JET is unresponsive to cardioversion and commonly used drugs like digoxin, beta blockers and calcium channel blockers. This stems from the underlying mechanism of enhanced automaticity. Hypothermia supresses cellular automaticity and was introduced for treatment of JET before amiodarone became widely accepted . It continues to be an important tool in management.[69] It is often used in combination with pharmacotherapy/pacing.[32,34,36,61,70-73] In normal subjects, hypothermia reduces cardiac output and oxygen demand and increases systemic vascular resistance. Hence, oxygen supply/demand is unaffected, provided shivering response is abolished by paralysis and mechanical ventilation.[71,74] Accordingly, all cooling protocols require sedation, paralysis, and mechanical ventilation. Hypothermia is induced by application of ice packs, cooling blankets, fans, lavage using cold physiological saline through nasogastric tube or by intravenous induction using cold saline.[34,73] Cooling is usually done to a core temperature (rectal) of 32°C–34°C for 24–36 h. Lesser degrees of hypothermia can be used in milder cases. Re-warming is started once the patient is stable for at least 24 h. Resolution of arrhythmia is not the criterion for re-warming. Hypothermia helps in reducing the heart rate effectively such that the restoration of AV synchrony by pacing is possible.[36,72,73,74] Although hypothermia is known to suppress immune response, none of the studies reported an increase in infections; neither did they report metabolic acidosis nor electrolyte imbalances attributable to hypothermia.[34] Hypothermia is a safe and effective method for reducing tachycardia rate adequate enough to improve hemodynamics.

Pharmacotherapy

Minimal negative inotropy and success in congenital JET led to use of amiodarone in postoperative setting.[54,57] A randomized double blind study of amiodarone in pediatric population, 37% of which were post operative patients, half of whom had JET demonstrated the efficacy as well as safety concerns of amiodarone.[56] Dose-related adverse events included hypotension (36%), vomiting (20%), bradycardia (20%), AV block (15%), and nausea (10%).[56] Two postoperative deaths were supposed to be related to the drug as well. Other nonrandomized studies showed better efficacy and safety profile and amiodarone has emerged as the first-line agent.[35,54,56,57,75-77] Hypotension is frequent and is usually responsive to volume expansion or calcium. Increase in inotrope dose may be required. Variation in dosing of amiodarone is a common practice to counter this, often omitting the loading dose and using a slow infusion.[59] High AV oxygen saturation difference was an independent, significant risk factor for amiodarone failure and need of escalation therapy.[35]

Procainamide, a class IA anti-arrhythmic, has also been used successfully.[50,78] The active metabolite N-acetyl procainamide has class III antiarrhythmic properties. A higher bolus dose and a faster uptitration resulted in more effective control.[78] Blood pressure remained stable or increased and central venous pressure remained stable or decreased in all patients. There was no mortality related to JET in this study.[78] Hypotension is supposed to occur from ganglionic blockade at higher plasma concentrations, rather than negative inotropic effect which is minimal. Procainamide requires blood level monitoring.

Dexmedetomidine has a potential role in the acute phase of perioperative atrial and junctional tachyarrhythmias for heart rate control and conversion to sinus rhythm. Incidence of adverse events is 28%, mostly hypotension and rarely AV block.[79] In a more recent analysis of children undergoing surgical closure of ventricular septal defect, preincision initiation of dexmedetomidine, and continuation in postoperative period significantly decreased JET, as opposed to postoperative use.[80] Nifekalant hydrochloride, a class III anti-arrhythmic approved in Japan, has been reported to be useful in case reports.[81]

Newer agents

Sotalol has class III anti-arrhythmic properties similar to amiodarone and N-acetyl procainamide. It has been reported to be useful in intravenous form in a dose of 1 mg per kg over 1 h for postoperative JET.[82] Sotalol does not require monitoring of plasma levels. The side effect and performance profile compared to amiodarone and procainamide need to be ascertained.

The latest tool in the armamentarium is ivabradine.[83-85] Ivabradine is recommended for select adults with chronic stable angina, heart failure, and inappropriate sinus tachycardia. Ivabradine acts through HCN channels which may play an important role in etiopathogenesis. The major advantage of ivabradine, particularly in the postoperative patient, is its hemodynamic profile, where heart rate reduction is achieved without modifying inotropy, lusitropy, and intracardiac conduction as

opposed to other drugs.[86] In small case series, ivabradine has been found to be effective as an adjunct to amiodarone and as the sole agent.[83-85] Rate control is the initial response, followed by conversion to sinus rhythm.[84] Early administration before hemodynamic compromise ensues will be key as absorption will be affected once cardiac output is compromised. These promising results warrant larger, randomized studies comparing it to amiodarone.

Digoxin, beta-blockers, and calcium channel blockers are classically considered ineffective for JET.[32,36,61,62,70] Class IC agents – propafenone and flecanide – were also shown to be effective.[62,63,87,88] However, in addition to nonavailability of intravenous formulations, negative inotropic effects and proarrhythmic potential limit their use.

SUMMARY

JET is a common arrhythmia in the pediatric cardiac postoperative setting, arising from enhanced automaticity of His bundle. Multiple factors have been found to be associated. Diagnosis is usually made on surface ECG; atrial electrograms are helpful in diagnostic dilemma. JET has devastating effects on postoperative hemodynamics. Prompt recognition and immediate institution of appropriate treatment (and avoidance of treatments like DC shock which is ineffective and likely counterproductive) are important. In addition to general measures such as correction of fever, electrolyte abnormalities, anemia and hypovolemia, optimizing sedation and pain control, and reducing inotropic drugs, therapeutic hypothermia and pharmacotherapy help controlling JET. Amiodarone is currently the preferred pharmacologic agent; ivabradine appears promising. Pacing restores AV synchrony augmenting CO. Recognition of the negative effect of JET on postoperative hemodynamics and advances in management have significantly reduced mortality and morbidity. Large multicenter randomized controlled trials comparing pharmacotherapy, hypothermia, and combined pharmacotherapy and hypothermia with restoration of AV synchrony by pacing in all arms as soon as rate allows are needed. Considering that JET has a high rate of spontaneous resolution, and that success rates across various modalities are similar, time to JET resolution should be considered as a better outcome variable along with response rates in future studies.

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Conflicts of interest

There are no conflicts of interest.

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