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Guidelines for the management of junctional ectopic tachycardia following cardiac surgery in children

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Pacing

Summary
Junctional ectopic tachycardia (JET) is a relatively common arrhythmia following open-heart surgery, especially surgery involving the bundle of His. It may be associated with significant impairment of cardiac output and has been associated with increased mortality. Effective management involves identification of high-risk cases, preventative measures and early diagnosis. Treatment with surface cooling, anti-arrhythmic drugs (in particular, amiodarone) and external cardiac pacing have proven effective in restoring cardiac output in children with JET post-operatively.

Practice points

- Junctional ectopic tachycardia (JET) is the most common arrhythmia in children following cardiac surgery
- High-risk patients can be identified preoperatively
- Early recognition of JET is essential in reducing adverse consequences
- Surface cooling and amiodarone have been shown to effectively reduce the rate of JET
- Establishing external cardiac pacing will usually restore atrial–ventricular (AV) synchrony

Introduction

Junctional ectopic tachycardia (JET) is a relatively common arrhythmia following open-heart surgery (1–2% of all open-heart cases). Although it may occur rarely in a congenital idiopathic form it is most commonly seen following surgery involving the region of the bundle of His. Repair of tetralogy of Fallot, ventricular septal defect (VSD) and atrioventricular septal defect (AVSD), atrial partition for TGA (Mustard/Senning), lateral tunnel Fontan palliation and repair of total anomalous pulmonary venous return (TAPVR) are the procedures most commonly associated with JET. It has, however, been reported in extracardiac procedures such as extracardiac Fontan palliation, coarctation repair and pulmonary artery banding. It is more commonly seen in infants and often associated with transient AV block immediately post-cardiopulmonary bypass.[1] It is also associated with residual cardiac defects post-operatively in approximately 10–15% of cases.

JET is caused by abnormal automaticity of the AV node with AV dissociation. Autopsy reports of children with fatal JET have shown haemorrhagic tracks invading the AV bundle

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originating from sutures close to conduction system.[2] The morphology of the QRS is similar to that in sinus rhythm, although associated incomplete or complete bundle branch block may result in a wide QRS complex. AV dissociation may be difficult to confirm, particularly when there is retrograde 1:1 conduction of p-waves or p-waves are hidden in QRS complex. Atrial wire electrocardiogram ECG is usually confirmatory.[3]

JET usually appears within the first 72 h post-operatively and usually resolves spontaneously provided haemodynamic stability can be achieved. The heart rate is usually 160–260 beats/min with haemodynamic instability more common at higher rates. JET is often described as 'warming-up', i.e., the onset is insidious and gradual as opposed to re-entrant tachycardias, which are sudden in onset. JET may result in significant compromise to cardiac output due to loss of AV synchrony and ventricular filling in the heart already compromised following surgery and cardiopulmonary bypass.

There are several approaches to the management of JET comprising physiological alterations, drugs, external cardiac pacing, and extracorporeal support.

Increased adrenergic tone accelerates JET rate and may contribute to cardiac compromise, which in turn results in an increase in adrenergic activity, creating a vicious cycle. Thus avoidance or reduction of adrenergic (e.g. catecholamines) or vagolytic drugs (e.g. pancuronium) is the first goal. Correction of electrolyte and acid–base abnormalities, correction of fever and optimisation of sedation are also thought to beneficial. Following this there are a number of different treatment strategies.

Treatment strategies

Cooling

Induction of moderate hypothermia reduces the rate of automatic depolarisation of all cardiac cells and has the effect of slowing the rate of JET, thus improving haemodynamic status or allowing external cardiac pacing. Achieving temperatures of 33–35 °C with a cooling blanket, ensuring sedation and muscle relaxation (to prevent shivering) has been shown to reduce rate of JET[1,4] and also reduces metabolic demand. Complications related to cooling include an increase in systemic vascular resistance, decreased cardiac contractility, difficulties assessing haemodynamic status clinically, impaired immune function, coagulation abnormalities and delay in extubation and discharge from the intensive care unit (ICU).

Magnesium

Intravenous magnesium sulphate (30–50 mg/kg) has been shown to be effective in preventing JET post-operatively[5] and may be effective in treating established JET. Adverse effects include vasodilatation and hypotension. Infusion to maintain the serum level at 1.5–2.5 mmol/l may sometimes be required.

Anti-arrhythmic drugs

Procainamide (Type Ia)

A procainamide bolus of 5–15 mg/kg over 15–30 min and infusion of 20–120 μg/kg/min has been shown to be effective in controlling JET.[1,6] Adverse effects include proarrhythmic effects, bradycardia and hypotension.

Amiodarone (Type III)

Amiodarone causes prolongation of the action potential in atrial, nodal and ventricular myocardial cells. It also increases the repolarisation and refractory period and has non-competitive alpha- and beta-adrenergic effects. It has been shown to be effective in either controlling JET rate or reverting JET in at least 80–90% of cases.[7,8,12] It is given as a bolus, due to its wide volume of distribution, followed by infusion of 5–15 μg/kg/min. Although associated with a wide range of adverse events when used long-term, its acute side effects are mainly pro-arrhythmic effects (especially torsades de pointe), negative inotropy and hypotension due to calcium channel blockade. Hypotension may be treated with 10% calcium gluconate (0.5 ml/kg). Late sinus bradycardia has also been reported following cessation of amiodarone.[9]

Propafenone (Type 1c)

Propafenone has been shown to be effective in the management of JET,[10] but is not widely available outside North America. Adverse effects include pro-arrhythmia (especially torsades de pointe) and negative inotropy.

Beta-blocking agents (Type II)

Esmolol is a short-acting beta-blocking agent, which may be used to increase AV block and improve haemodynamic status. It has been used effectively, particularly in late JET. Sotalol has also been used with some effectiveness in reducing JET rate.[11]

Digoxin

Once a mainstay of treatment, digoxin has been used less frequently in recent years. In a large study from Boston Children's Hospital evaluating a treatment protocol it was found to be ineffective.[1] Its adverse effects include pro-arrhythmic effects.

External cardiac pacing

The main objective of the above measures is to slow the rate of JET. Haemodynamic problems are commonly encountered when the rate is greater than 170 beats/min. If cardiac output is not improved by slowing the JET rate, external cardiac pacing may be useful in attaining AV synchrony, improving left ventricular filling and cardiac output. External cardiac pacing is usually successful at a rate slightly above the underlying JET rate. Atrial pacing (AAI) may be successful,[9] but if there are problems with AV conduction related to underlying process or anti-arrhythmic agents, AV synchronous pacing (DDI) may be required.[13] Overall the goal is optimal AV synchrony. Alternative pacing manoeuvres such as paired ventricular pacing may be effective in improving cardiac output.[14]

Radiofrequency or cryoablation

Both emergency cryoablation[15] and radiofrequency ablation[16] have been used successfully in severe, intractable JET. Long-term AV conduction may be better preserved with radiofrequency ablation.

Mechanical support

Mechanical support such as left ventricular assist device (LVAD) or extracorporeal membrane oxygenation (ECMO) may be effective as rescue therapy in severe, intractable JET.[17]

Guidelines for the management of JET

Prophylaxis

It is recommended that all patients at increased risk be given 50 mg/kg magnesium sulphate immediately after cardiopulmonary bypass before transfer from theatre. Normothermia should also be ensured post-operatively with surface cooling if necessary. Those at high risk include infants having undergone repair of tetralogy of Fallot, VSD, transposition with VSD, AVSD, lateral tunnel Fontan palliation and repair of TAPVR.

Diagnosis

The ECG-rate is usually 180–250 beats/min and regular. The QRS is usually narrow complex and p-waves may be hidden, dissociated or retrograde. Diagnosis should be suspected in those cases in which JET is common, i.e., Fallot's, VSD, AVSD. Atrial ECG is usually confirmatory in difficult cases. In comparing atrial ECG with surface ECG, atrio-ventricular dissociation can be elegantly demonstrated (Fig. 1a and b).

Assessment of haemodynamic stability

Haemodynamic instability or reduction in cardiac output becomes more likely at rapid JET rates (>170/min). It is important to ascertain whether cardiac output is significantly affected. Signs of haemodynamic instability are listed in Table 1.

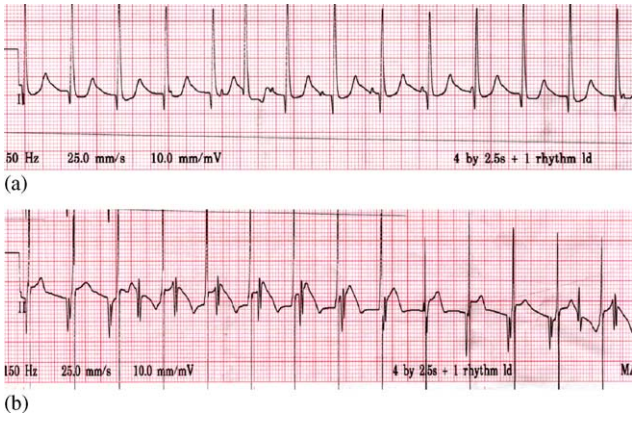

Figure 1 (a) Junctional ectopic tachycardia demonstrating atrio-ventricular dissociation. (b) Atrial ECG (wire study) demonstrating large atrial (p-wave) spikes.

Haemodynamically stable JET

When the patient with JET is haemodynamically stable, it is essential to maintain vigilance for any increase in rate or any indication of haemodynamic compromise. Electrolyte and blood gas abnormalities should be promptly corrected. Avoidance or minimisation of drugs that may drive the JET rate is also important. These drugs include sympathomimetics (e.g., catecholamines) and vagolytic drugs (e.g., pancuronium). Fever will also increase the underlying rate of JET and all febrile patients should be cooled to normothermia (36.5 °C) with surface cooling. Adequate sedation and analgesia is also important. Most importantly, continual assessment of haemodynamic status and cardiac output is mandatory. This includes frequent clinical examination, serum lactate and mixed venous oxygen saturation. More invasive assessment of cardiac output such as echocardiography, thermodilution techniques and Doppler studies may provide additional information. The use of antiarrhythmics such as amiodarone in the stable patient is controversial. Although amiodarone may control JET rate and allow overdrive pacing, potential adverse effects need to be taken into account in the stable patient.

Haemodynamically unstable JET

When haemodynamic compromise becomes evident or there are indications that cardiac output is compromised, treatment needs to immediate. The above measures all need to be continued, but additional treatment is required (Table 2).

Resistant/intractable

In many instances, JET may be intractable to standard treatment. There a number of second-tier treatment options, which may be effective. A further reduction in temperature to 32–33 °C with surface cooling may be applied. This may further slow the rate of tachycardia. At lower temperatures, however, adverse effects are more likely to be seen. Increasing the rate of amiodarone up to 15 μg/kg/min is another option. Other antiarrhythmics may also be considered, such as procainamide, propafenone,

Table 1 Signs of haemodynamic instability.

Hypotension
Poor peripheral perfusion
Significant inotrope requirement
Escalation of inotrope requirements
Oliguria (urine output < 1 ml/kg/h)
Rising serum lactate
Metabolic acidosis
Decreased mixed venous oxygen (< 60%)
Increased core-skin temperature gradient (> 5 °C)

Table 2 Interventions for haemodynamically unstable JET.

Interventions	
Amiodarone	Bolus 5 mg/kg Infusion 5–10 μg/kg/min Beware of hypotension/ bradycardia
Active cooling	34–35 °C Ensure adequate sedation/ muscle relaxation
Magnesium sulphate	50 mg/kg over 20–30 min Maintain serum level 1.5–2.5 mmol/l
External cardiac pacing	Initiate at 10 beats/min above JET rate Atrial pacing (AAI) preferred to dual (DDD)
Reassess echocardiography	Repeat transoesophageal or transthoracic

flecainide and esmolol. Mechanical support, either LVAD or ECMO, should be considered in those patients who have intractable JET with cardiac compromise despite the above measures.

Weaning

Once patient has reverted to sinus rhythm, weaning from interventions can be considered. Weaning should occur in a stepwise fashion. Temperature should be gradually returned to normal with reduction in sedation and muscle relaxation. Slow reduction in infusion rate of amiodarone should then be attempted. As appropriate the rate of infusion should be halved and then stopped. The half-life of amiodarone may be prolonged especially if haemodynamic compromise has been present or infusion has been prolonged. Late sinus bradycardia secondary to amiodarone may occur and should be monitored.

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