



Question of the Week

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Keyword	Junctional ectopic tachycardia
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Question

A 5-month-old male with a history of Tetralogy of Fallot (TOF) underwent complete repair with transannular patch and VSD closure. On arrival to the intensive care unit, the ECG shows narrow complex tachycardia with AV dissociation and heart rates between 170-210 beats per minute (bpm), with a systolic blood pressure in the 60s. Axillary temperature is 35.4°C and most recent labs are within normal limits, with a potassium of 4.2 and magnesium 2.7. An amiodarone infusion is started after a loading dose. Which of the following is the BEST NEXT course of action?

- A. Atrioventricular sequential pacing
- B. 30mg/kg IV magnesium sulfate
- C. Consult electrophysiology (EP) team for catheter ablation

Explanation

Junctional ectopic tachycardia (JET) is a rare arrhythmia often seen in patients with congenital cardiac disease, originating from the atrioventricular (AV) junction or bundle of His¹. JET is characterized by abnormal automaticity rather than reentrant mechanisms. It presents as a narrow complex tachycardia with AV dissociation, often at rates of 200-250bpm¹. ECG findings include gradual acceleration and deceleration, distinguishing it from other tachyarrhythmias. Hemodynamic compromise is common due to rapid ventricular rates and loss of synchronized atrial contraction, resulting in decreased cardiac output.

There are two types of JET: congenital and post-operative. Congenital JET presents without a history of cardiac surgery with etiologies ranging from genetic predisposition, intrinsic abnormalities of the conduction system, ion channel dysfunction or histopathologic changes such as AV nodal fibrosis. Congenital JET tends to be refractory to treatment, increasing the risk of heart failure and sudden cardiac death. Mortality has been reported as high as 35% in untreated or refractory cases of congenital JET. Post-operative JET typically occurs within the first 72 hours following congenital cardiac surgery³. Mechanisms for post-op JET include ischemia, mechanical stretching or trauma to the AV conduction system. Certain anatomical features and surgical procedures increase the risk, with those that involve structures near the AV node having the highest risk. Although post-operative JET is self-limited and will resolve spontaneously within 5-7 days, it can lead to significant morbidity and mortality³.

A summary of risk factors that increase the risk of post-operative JET¹:

1. Type of procedure (Repair of TOF, double-outlet right ventricle (DORV), ventricular septal defect (VSD), atrioventricular septal defect (AVSD), transposition of the great vessels)
2. Age < 6 months
3. Higher post-operative core temperature
4. Use of inotropes (epinephrine, milrinone, dopamine)
5. Electrolyte abnormalities (hypokalemia, hypomagnesemia)
6. Prolonged cardiopulmonary bypass times (≥ 75 minutes)

The three main goals of managing JET are: rate control, restoring AV synchrony, and treatment of any underlying causes. Treatment focuses on correcting electrolyte abnormalities (specifically potassium, magnesium and calcium) administering antiarrhythmics, sedation, and cooling³. In refractory cases, catheter-directed ablation can be utilized. Although adenosine will not terminate JET, it may slow down the rhythm enough to confirm the absence of p waves, helping to distinguish it from other tachyarrhythmias.

Treatment of JET includes the following¹⁻⁴:

1. Replace potassium, magnesium, and calcium as needed
2. Correct acid-base status (especially metabolic acidosis)
3. Decrease inotropic agents if appropriate
4. Avoid/treat fever and consider instituting mild therapeutic hypothermia (34-35 degrees)
5. Amiodarone (loading dose: 5mg/kg over 30 minutes or slower, infusion 10-20mg/kg/day)
6. Consider sedation (i.e. dexmedetomidine) to reduce sympathetic tone
7. Pacing to restore AV synchrony

Studies have found other common agents used in anesthetic practice may be helpful in management, such as dexmedetomidine. In a recent study, prophylactic dexmedetomidine significantly lowered the rate of postoperative JET compared to placebo (3.3% versus 16.7%)⁴. Pacing can restore AV synchrony, suppressing the junctional automaticity, allowing for improvement in hemodynamics¹.

Across major pediatric cardiac critical care centers, both medication and pacing are the most commonly utilized treatments. It is important to note that there may be significant variation in institutional practices for JET management. For example, although amiodarone is widely considered the first-line drug⁵, other medications, such as procainamide, are also used successfully. Cooling was used less frequently and typically in conjunction with other therapies, rarely as the sole management strategy⁴.

The correct answer is choice A, AV sequential pacing. The serum magnesium is within normal limits, and other treatment modalities are warranted prior to considering catheter-directed ablation for post-operative JET.

References

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Answer

- A. Atrioventricular sequential pacing