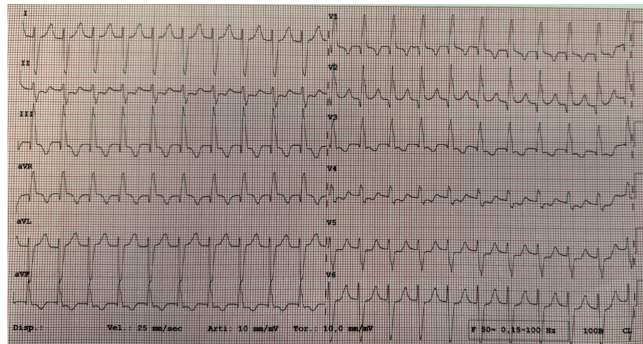


Palpitations in a Fontan patient: what now?

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A 32-year-old male presented to our emergency department with palpitations. His blood pressure was 95/65 mmHg, heart rate 150 beats per minute, oxygen saturation 87% on room air, and body temperature 36°C. The electrocardiogram (EKG) showed paroxysmal supraventricular tachycardia (PSVT). He reported a history of heart surgery – Fontan procedure¹ – during childhood for a complex congenital heart disease (CHD) characterized by a single atrium, a single ventricle, and a single atrioventricular valve. A focused cardiac ultrasound confirmed a single atrium, a single ventricle, no pericardial effusion, and an estimated ejection fraction of 50% (Videos 1 and 2). His current medication regimen included sotalol 80 mg, losartan 12.5 mg, dapagliflozin 10 mg, furosemide 25 mg, and warfarin.

Question

Considering the patient's medical history, which medication is the most appropriate choice?

1. Adenosine.
2. Non-dihydropyridine calcium channel blockers (Class IV: diltiazem or verapamil).
3. Beta-blockers (Class II: metoprolol or esmolol).
4. Electric cardioversion.

Answer

Managing PSVT can be particularly challenging due to the significant variability in the condition and the evolving nature of adult CHD. There is currently no randomized controlled trial data available to guide pharmacological management. Atrial arrhythmias are a well-recognized and common complication associated with Fontan palliation, occurring in approximately 15-60% of patients, with the incidence increasing with age.² These arrhythmias may result from factors such as suture lines, injury to the sinus node, compromised arterial supply, and atrial remodeling.³ Recommendations are primarily based on expert consensus and

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Informed consent: the patient provided informed consent for access to medical records at the time of admission.

Availability of data and materials: all data underlying the findings are fully available upon reasonable request to the corresponding author.

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retrospective studies, as well as a recent position paper from the European Heart Rhythm Association (EHRA) regarding the diagnosis and treatment of arrhythmias in patients with CHD. While the general approach aligns with standard guidelines, it necessitates specific considerations for this unique patient population. Adenosine is the treatment of choice for the acute management of atrioventricular nodal-dependent mechanisms. However, its use is not recommended in patients with reduced ejection fraction who rely on high heart rates to preserve cardiac output, and caution is warranted in cases of sinus node dysfunction.^{4,5} Negative inotropic agents such as beta-blockers and non-dihydropyridine calcium channel blockers can lead to cardiovascular collapse in patients with compromised ventricular function. Therefore, an echocardi-

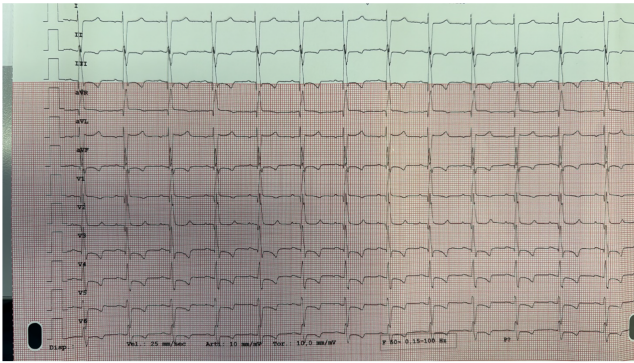


Figure 1. EKG showing sinus rhythm.

graphic assessment is crucial prior to administering these medications, which should be administered via slow infusion rather than bolus injection to minimize risks.⁵ All patients with hemodynamically unstable supraventricular tachycardia require urgent electrical cardioversion.⁶ Our patient was initially treated unsuccessfully with modified Valsalva maneuvers. Subsequently, diltiazem at a dose of 30 mg was infused over approximately 10 minutes, resulting in the restoration of sinus rhythm (66 bpm), blood pressure of 100/60 mmHg, and SaO₂ of 93%. The EKG showed a sinus rhythm with T-wave inversion, consistent with previous EKG findings (Figure 1). After a few hours, the patient was discharged without any symptoms and referred to his cardiology center for follow-up.

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Online Supplementary Materials:
Video 1. Fontan heart.
Video 2. Apical 2-chamber Fontan.