# Cardiac pacing in patients with Fontan circulation: Further considerations. Authors' reply

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We very much appreciate your careful reading and thoughtful response to our article "Atrioventricular sequential pacemaker implantation in an adult patient with Fontan circulation"[1]. We agree that it is challenging to accomplish permanent cardiac pacing in patients with Fontan circulation (FC) due to complex anatomy, electrophysiologic abnormalities, and limited access to cardiac chambers. Mindful of the risks related to a high percentage of ventricular pacing in this specific group of patients, the final pacing program of the Vitatron G20A2 DR pacemaker was set to DDD pacing mode with a preference for endogenous conduction utilizing the AV search algorithm with a prolongated AV interval [2]. The ventricular lead was implanted to serve as a backup for intermittent atrioventricular (AV) conduction disturbances observed in previous ECG tapes. During follow-up, the percentage of ventricular pacing due to episodes of sino-atrial dissociation or AV block was <10% in pacemaker histograms. The pacing parameters remained stable, and the systolic function of the systemic ventricle did not change.

The presence of fenestration in FC is known to have advantages in the immediate postoperative course and long-term outcomes comparable to a non-fenestrated approach. In addition, patients with preserved Fontan fenestration have a stable hemodynamic response with a secured preload reserve, reduced afterload, suppressed beta-adrenergic response, and lower baseline HR in stress tests [3]. Furthermore, an additional role of fenestration in FC is to reduce venous stasis, thereby reducing passive congestion of the liver. Our previous research showed that chronotropic failure was associated with multiple organ damage, including liver dysfunction, and improving the chronotropic capacity reduced passive congestion of the liver [4].

Fenestration in FC is the only route that enables advance of the pacing leads into the heart. However, pacemaker leads reduce the fenestration area, which might negatively impact the patient's exercise capacity. In the case of a small fenestration, it is technically possible to dilate it with angioplasty to fit in the pacing leads and preserve the blood flow through the fenestration [5]. The authors have planned a regular follow-up in an adult congenital heart disease clinic to ascertain the patient's favorable long-term outcomes, including echocardiography, exercise tolerance tests, and, if required, FC catheterization with hemodynamic measurements. In conclusion, the authors agree that cardiac pacing implications in the FC setting are still to be established.

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