

## Cardiac pacing in patients with Fontan circulation: Further considerations

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Fontan circulation (FC) denotes a direct connection between systemic venous return and the pulmonary arterial system and is universally created through a variety of palliative surgical techniques (namely the Fontan procedure) in patients with complex congenital anomalies such as tricuspid atresia [1–3]. In particular, patients with FC might encounter a variety of complications including arrhythmias, systemic ventricular failure, etc. [2, 3]. In their recently published clinical vignette, Boczar et al. [1] have reported a challenging case of dual-chamber pacemaker implantation in the setting of FC. Therefore, we would like to comment on this interesting case and its implications.

First, a portion of patients with FC might suffer progressive ‘systemic ventricular failure’ due to a variety of factors such as reduced ventricular preload and might require radical operations, including heart transplantation, in the long term [2]. In clinical practice, cardiac resynchronization therapy (CRT) (with variable responses) has also been performed in patients with decompensated FC, who might be more likely to harbor mechanical ventricular dyssynchrony (compared with those having biventricular circulation) [4], possibly in association with impaired ventricular conduction as demonstrated with a significant QRS prolongation. However, ventricular pacing in FC patients with intact ventricular conduction (performed for bradyarrhythmias) [2] might, on its own, lead to a dyssynchronous contraction pattern (presenting with relative increases in QRS duration) along with potential disturbances in atrioventricular (AV) synchrony [4]. Importantly, these pacing-related alterations in patients with FC might have a significant impact on the evolution and/or

aggravation of ‘systemic ventricular failure’ in the long term. Accordingly, ventricular pacing in these patients was previously reported to have a significant adverse impact on cardiac output and quality of life, possibly leading to eventual death and cardiac transplantation [2, 4]. Notably, atrial pacing is devoid of these adverse effects (as this mode of pacing potentially simulates physiological cardiac automaticity and conduction) [4].

In line with the above-mentioned notions, the normal ventricular conduction pattern in the patient was found to be relatively impaired (resulting in significant QRS prolongation) following DDD (dual-chamber) pacing [1]. Accordingly, we wonder about systemic ventricular functions of the patient (before pacemaker implantation and on follow-up). Was she completely pacemaker-dependent? Substantial preponderance of ventricular-paced beats (a ratio of  $\geq 50\%$ ) might pose a significant challenge in terms of ‘systemic ventricular failure’ and associated conditions [4]. Therefore, since the AV node seems unaffected [1], it would be reasonable to switch the patient to atrial pace-sense mode (AAIR). In particular, an electrophysiological study (EPS) evaluating AV node functions might help determine the need to switch back to DDD pacing, where necessary, in the future.

Finally, fenestration in the Fontan baffles reduces systemic venous pressure and contributes to the maintenance of systemic output due to reverse shunting, particularly during exertion [2]. Accordingly, reduced fenestration area (due to pacemaker leads) [1] might have a negative impact on the patient’s exercise capacity [2] (despite her improved chronotropic response). Therefore, the patient [1] needs to be more frequently examined

(with regard to systemic venous congestion and blood pressure response to exercise) for the timely initiation of further therapeutic strategies [2]. Therefore, what would be the authors' follow-up strategy for their patient [1]?

In conclusion, the authors [1] did an excellent job for their patient, in whom surgical pacemaker implantation might be quite challenging (due to adhesions from previous recurrent surgeries) [4] and possibly more risky. Importantly, cardiac pacemaker implantation and its indications (for resynchronization or bradyarrhythmias), along with modes of cardiac pacing (AAI versus DDD, etc.), might significantly affect the cardiovascular status of patients with FC [2, 4] due to their limited physiological reserve. However, further implications of cardiac pacing in the setting of FC still need to be established.

### Article information

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