Ablate and pace as bail-out therapy in a patient with Fontan correction and malignant atrial tachycardia

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Atrial tachyarhythmias complicating Fontan correction may have a ‘malignant’ clinical presentation seriously impairing the patient haemodynamic. Current strategies are surgical total cavopulmonary connection with or without antiarrhythmic surgery or transcatheter ablation. We describe the case of a patient who previously underwent atroiovacal Fontan correction and later presented with refractory atrial tachycardia responsible for relapsing syncope. After a failed attempt at surgical conversion, and while waiting for heart transplantation, he was submitted to ablation of the atrioventricular node through an aortic retrograde approach and ventricular pacing through the coronary sinus tree. One year later, the patient is doing well, displaying a stable functional recovery and excellent pacing lead performances. An ablate and pace approach may deserve consideration in selected Fontan patients experiencing life-threatening atrial arrhythmias.

Introduction
Atrial arrhythmias are common following Fontan correction of tricuspid atresia and their incidence increase with the post-operative interval.1,2 At least 50% of patients experience clinically significant atrial arrhythmias during an average 20-year follow-up. These arrhythmias are mostly represented by intra-atrial re-entrant tachycardias (IARTs) and atrial fibrillation.3–6 These arrhythmias may be due to both lines of block deployed during previous surgical intervention, and atrial structural and electrical remodelling.7 The occurrence of such arrhythmias significantly impairs patient survival, prompting an immediate action in order to avoid their detrimental consequences. Antiarrhythmic therapy alone is poorly effective and limited by side effects prompting dose reduction over time. Alternative therapeutic options proposed so far are surgical conversion to total cavopulmonary connection with atrial debulking or percutaneous radiofrequency ablation with electroanatomic mapping. The former has become the treatment of choice when combined with MAZE, although burdened by a not negligible procedural risk,8 the latter displays both a variable acute success rate and a high percentage of recurrence at mid-term follow-up (up to 50% at 2 years).9 When both options are technically unfeasible or unsuccessful, heart transplantation can be considered as a last resort, although associated with a high early mortality.10

We are describing an alternative effective strategy chosen in a patient with significant haemodynamic impairment and relapsing syncope due to refractory atrial tachycardia.

Case report
A 29-year-old male patient who had undergone extracardiac Fontan correction owing to pulmonary atresia was proposed for a bail-out attempt of ablation after recurrent syncope due to atrial tachycardia (IART).

At birth, he was diagnosed with pulmonary atresia associated with dextrocardia—situs viscerus solitus. At the age of 5, he underwent the Fontan operation—Kreutzer modified procedure.11 Following the palliation surgery, he did well for a decade. At the age of 15, he had several syncopal episodes due to IART rapidly conducted to the ventricle. Arrhythmias were controlled with amiodarone until the age of 25 when syncopal IART relapsed (Figure 1). In 2003, myocardial nuclear scintigraphy showed mild ventricular insufficiency (ejection fraction 45%). Amiodarone was given up due to iatrogenic hyperthyroidism and sotalol was started with poor benefit. At the age of 28, an attempt of conversion to total cavopulmonary connection was suspended after sternotomy due to cardiac unfavourable anatomy which exposed the patient to a very high procedural risk. Taking into consideration the progression of the symptoms, the patient was scheduled for atrioventricular (AV) ablation and endocardial pacemaker implantation (‘ablate and pace’ approach).12 Two active fixation leads (Starfix®, Medtronic) were implanted in two postero-lateral branches of the coronary sinus and were connected to a biventricular pacemaker (InSyncIII®, Medtronic) (Figure 2).

The Attain Starfix LV lead, model 4195, is a 5 Fr, steroid-eluting, unipolar, polyurethane insulated cardiac vein lead that features deployable lobes to provide fixation. In the same procedure, the His bundle was ablated with a retrograde aortic approach, owing to inaccessibility through the venous system.
At 3-month follow-up, the patient was doing well, his clinical condition improved to such an extent that he was removed from the transplantation waiting list.

After 1 year, the patient is still asymptomatic; pacing impedance and threshold are stable and the percentage of pacing is permanently 100% over 24 h.

**Discussion**

It is well recognized that supraventricular arrhythmias in patient with congenital heart disease may lead to a critical haemodynamic impairment, usually prompting an immediate treatment.

**Figure 1** Episode of symptomatic IART during telemetric monitoring in the intensive care unit where the patient was admitted for recurrent syncopal episodes. The three strips are consecutive and show an ECG tracing (upper) and the intra-arterial blood pressure recording (bottom). Note the haemodynamic impairment during fast conduction of a IART episode (strip A). As soon as the ventricular rate slows due to an increase in the degree of AV block (strip B), arterial blood pressure significantly improves. After interruption of IART, sinus rhythm competes with junctional rhythm while the arterial blood pressure returns to normal value (strip C).

**Figure 2** Selective venography in the right anterior oblique (RAO) projection (30°) of the coronary sinus tree showing a huge posterolateral vessel that divides into two branches near the free wall (A). Two Starfix leads were implanted in these two branches and their position is shown in the same RAO projection (B).
As far as the clinical setting of Fontan correction is concerned, arrhythmias are mostly represented by macroreentrant atrial tachycardia and atrial fibrillation. As previously stated, among the possible therapeutic options, surgical Fontan extracardiac reconversion with atrial debulking and transcatheter ablation are those most extensively accepted. The former, in particular, has become the treatment of choice following the publication of Chicago results. Their rationale stems from the pathophysiological notion that preservation of a regular rhythm with AV synchronization is pivotal in stopping and even reverting the vicious circle responsible for progressive and in most cases irreversible haemodynamic impairment. Both approaches may have important drawbacks in terms of surgical risk and low success rate, respectively.

We propose an alternative strategy, i.e. ablation of the AV node and ventricular pacing, based on the hypothesis that rate control and regularization of RR intervals is the actual target to achieve in order to allow the haemodynamic recovery. In Fontan corrected patients, the right atrium becomes a huge chamber in direct continuity with the cava veins allowing retrograde flow; thus, it is conceivable that its haemodynamic contribution might play a trivial role. The loss of the atrial contribution to the filling of the systemic ventricle is of course the downside of this approach and must be weighed against the benefit of rhythm regularization and diastolic interval optimization.

Few technical issues have been addressed when deciding to schedule this patient for an ablate and pace strategy. First, the ventricular stimulation was performed through the coronary sinus tree, owing to the inaccessibility of the ventricle from the venous system. Secondly, two coronary leads were implanted to both improve the safety margin, since the patient is pacemaker-dependent, and optimize the mechanic of left ventricular stimulation. Furthermore, the particular anatomic arrangement of the patient made it impossible to target the AV node through the venous system, prompting a left-sided retrograde approach. Finally, the thorough evaluation of these patients must take into consideration the risk of sudden death from ventricular arrhythmogenesis that might eventually mandate implantable cardioverter defibrillator implantation. In our particular case, the patient did not meet any conventional class 1 indication; moreover, the implantation would have been clearly impossible through the conventional transvenous route.

This case provide some pathophysiological demonstration that rate control, no matter how it is achieved, might be regarded as the key strategy able to counteract the progressive and life-threatening haemodynamic impairment associated with this syndrome. The uneventful follow-up, as far as the efficiency of the stimulation and the contractile performance are concerned, might dissipate some concern about the safety of the trans-coronary sinus ventricular stimulation in pacemaker-dependent patients.

Conclusions
According to large experiences, total cavopulmonary conversion along with antiarrhythmic treatment aiming at preserving AV coordination should still regarded as a first-line approach, nevertheless from our experience ablate and pace might deserve consideration in selected Fontan patients (high surgical risk) experiencing life-threatening atrial arrhythmias, taking into account the low procedural risk and high success rate when compared with the traditional therapies proposed so far.

Conflict of interest: None declared.

References