Development of Tachyarrhythmias Late After the Fontan Procedure

The Role of Ablative Therapy

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KEYWORDS

• Ablative therapy • Fontan procedure • Tachyarrhythmia • Atrial • Ventricular

KEY POINTS

- Patients with a Fontan circulation seem to be at high risk of developing a variety of atrial tachyarrhythmias (ATs) and ventricular tachycardias (VTs) at a relatively young age.
- The mechanisms underlying AT are variable, including both ectopic activity and reentry.
- Over time, successive AT may be caused by different mechanisms.
- The acute success rate of ablative therapy for AT is considerably high, yet, during long-term follow-up, recurrences frequently occur.
- It is most likely that these recurrences are caused by a progressive atrial cardiomyopathy rather than arrhythmogenicity of prior ablative lesions.

INTRODUCTION

In 1971, Francis Fontan and colleagues[1,2] from Bordeaux were the first to report on a new operation for patients with a single functional ventricle. The initial procedure was aimed at redirecting systemic venous blood directly into the pulmonary circulation without passing through the subpulmonary ventricle by constructing a connection between the right atrium and pulmonary artery.[1,2] Ever since, numerous modifications have been made to this procedure. Patients with a Fontan circulation are at high risk for developing a variety of cardiac dysrhythmias after cardiac surgery.[3] These dysrhythmias are most often supraventricular tachyarrhythmias (SVTs) but ventricular tachyarrhythmias may occur as well.[4] The quality of life of Fontan patients with tachyarrhythmias is often seriously affected by recurrent symptoms, such as fatigue, palpitations, and syncope. In addition, they may cause severe complications, such as hemodynamic deterioration, thromboembolic events, and even sudden cardiac death. In Fontan patients who died of heart failure, approximately 90% had SVTs.[5] The morbidity and mortality associated with tachyarrhythmias in this patient group justify close follow-up and aggressive arrhythmia therapy.

INDICATIONS FOR THE FONTAN PROCEDURE

In patients with single ventricle physiology, the most commonly applied therapy is surgical palliation by a Fontan procedure.[1,2] In this regard, the term *Fontan procedure* includes all variants of circulation in which the systemic venous return passes through the pulmonary vascular bed without the driving force of a subpulmonary ventricle. Over time, several determinants for successful outcome have been defined.[6] Essentially, the most important determinant is the pulmonary vascular bed. Preferably, the pulmonary vascular

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bed should have normal anatomy and a low or normal vascular resistance and the systemic ventricle should have a good function with no regurgitation of the atrioventricular valves. In combination with sinus rhythm, this provides the best pull function of the systemic ventricle with limited increase of central venous (pulmonary arterial) pressure as the driving force of the pulmonary circulation.

DIFFERENT TYPES OF FONTAN PROCEDURES

Originally, Fontan and Baudet[1] described their operation for palliation of tricuspid atresia as a single-stage atriopulmonary or atrioventricular connection. Although a Fontan circulation with an atriopulmonary or atrioventricular connection may be initially successful, many patients developed complications during long-term follow-up. Most of all, this concerns progressive right atrial dilatation and consequently atrial tachyarrhythmias (ATs), resulting in a loss of atrial transport function and a further decrease of cardiac output in these patients.

The total cavo-pulmonary connection (TCPC) was introduced in an attempt to overcome this problem.[7] Designed as a single-stage operation, nowadays most surgical centers apply the staged approach, with a partial cavo-pulmonary connection (of which several variants exist) in the first year of life to divert the superior caval blood directly to the pulmonary circulation. The Fontan circulation is completed in a separate operation to a TCPC by directing the inferior caval blood to the pulmonary circulation as well.[8]

This procedure is accomplished by an intra-atrial lateral tunnel, which has the advantage of growth potential. However, it needs intracardiac surgery with extracorporal circulation. Fontan completion can also be done with an extracardiac conduit, which has the disadvantage of a fixed (sometimes gradually declining) diameter and may lead to suboptimal hemodynamics.

However, an extracardiac conduit does not require open-heart surgery or extracorporal circulation. Apart from the scars of previous surgical procedures and the closing line of the superior and inferior caval vein, no additional intra-atrial scars are introduced. Whether the extracardiac conduit approach truly reduces the incidence of late atrial arrhythmias is still a matter of debate.[9,10]

Atrial dilatation and supraventricular arrhythmias were increasingly observed during long-term follow-up of patients with an atriopulmonary and atrioventricular Fontan circulation.

One option for patients who develop significant hemodynamic or arrhythmic problems after a classic atriopulmonary or atrioventricular Fontan is to perform an operation to convert them to a TCPC, either with a lateral intra-atrial tunnel or with an extracardiac conduit.[11–15]

PATHOPHYSIOLOGY OF SUPRAVENTRICULAR TACHYARRHYTHMIAS

Tachyarrhythmias are generally caused by abnormalities in either impulse formation (ectopic activity caused by triggered activity or enhanced automaticity) or impulse conduction (reentry). In patients with a Fontan circulation, SVTs are most often ATs, including intra-atrial reentrant tachycardia (IART), type I typical atrial flutter (AFL) or atrial fibrillation (AF); focal atrial tachycardias (FATs) have been less frequently observed.[16,17] Atrioventricular (nodal) reentrant tachycardia has, to the authors' knowledge, only been described in case reports.[18,19] Risk factors for SVT in patients with a Fontan circulation include right atrial enlargement, elevated atrial pressure, dispersion of atrial refractoriness, sinus node dysfunction, older age at the time of cardiac surgery, elevation of pulmonary pressure, low oxygen saturation, preoperative arrhythmias, prior palliation with an atrial septectomy, atrioventricular valve replacement, and aging.[20–22] The type of surgical technique used may also be related with the incidence of SVT. In 1991, Balaji and colleagues[23] found a lower incidence of late postoperative AT in patients who underwent a total cavo-pulmonary connection compared with patients who had a direct atriopulmonary connection. This observation was attributed to the lower right atrial pressure and subsequent atrial wall distension in patients with a total cavo-pulmonary connection.

In patients with a Fontan circulation, precipitating factors for development of reentrant tachycardias are the presence of prosthetic materials, electropathologic alterations of the atrial myocardium and chronic bradycardia due to damage to the sinus node, the sinus node artery, or usage of β-blocking drugs.[4,24] The myocardium is further affected by an ongoing pressure/volume overload and progressive scarring along suture lines or atriotomies. These structural changes cause intra-atrial conduction delay and regional differences in dispersion of atrial refractoriness. Local conduction abnormalities combined with a higher incidence of premature atrial beats due to stretch of myocardial fibers make these patients more prone to development of SVT.

MACRO-REENTRANT TACHYCARDIAS

As demonstrated in the left panel of **Fig. 1**, endovascular mapping studies before ablation of AT in patients with a Fontan circulation described as peri-tricuspid, counterclockwise rotation of a