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CASE REPORT

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Initial syncope associated with alternating attacks of supraventricular tachycardia and atrioventricular block long after surgical correction of tricuspid atresia

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Abstract

The Fontan procedure has become a generic term to define a surgical procedure that orients the systemic venous return directly to the pulmonary arteries, and has been used as a palliative operation for pulmonary atresia, tricuspid atresia, double inlet ventricle and complex single ventricle.

The earliest type of Fontan procedure was a simple atriopulmonary anastomosis between the right atrium and the pulmonary artery. Atrial arrhythmias, particularly atrial flutter and sinus node dysfunction may occur in the early and late postoperative period after simple atriopulmonary anastomosis. The case presented here represents a much delayed occurrence of an initial syncope due to alternating attacks of SVT (supraventricular tachycardia) and second degree heart block on admission 21 years after simple atriopulmonary anastomosis performed for the correction of tricuspid atresia. (Cardiol J 2008; 15: 186–188)

Key words: tricuspid atresia, Fontan operation, syncope, atrial arrhythmia

Case report

A 35-year-old female patient was admitted to our centre in an unconscious state with a blood pressure of 60–45 mm Hg and a supraventricular tachycardia (SVT) attack on initial electrocardiogram (Fig. 1). Within a few minutes the tachycardia on the monitor suddenly converted to second degree atrioventricular block (Mobitz Type 2) (Fig. 2). Within a minute, the patient gained consciousness with the restoration of normal sinus rhythm (NSR). The symptoms of near-syncope had been present

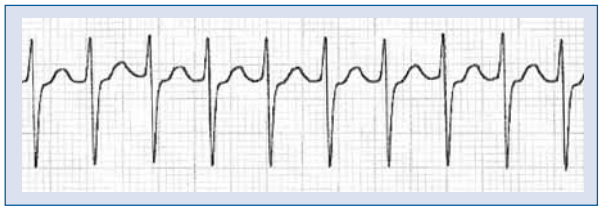

Figure 1. Supraventricular tachycardia on admission.

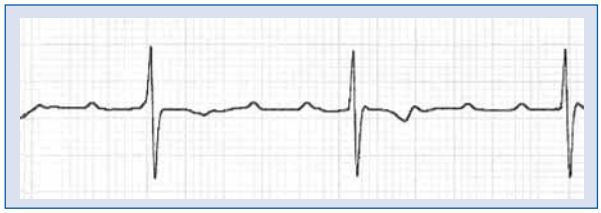

Figure 2. Spontaneous conversion to atrioventricular block.

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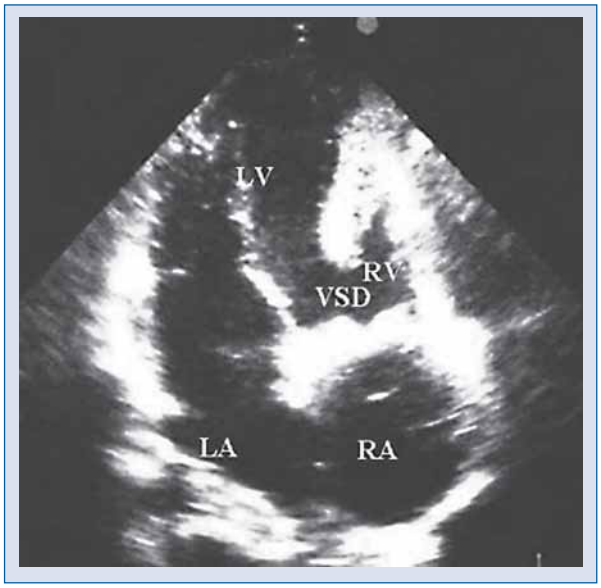

Figure 3. Transthoracic echocardiography demonstrating ventricular septal defect (VSD), hypoplastic right ventricle (RV) and atretic tricuspid valve (apical four-chamber view); LV — left ventricle; LA — left atrium; RA — right atrium.

for the last five days before admission. She had undergone Fontan operation (atriopulmonary anastomosis) 21 years ago for correction of tricuspid atresia. There was no history of any medication or co-morbid condition including coronary artery disease. The patient did not have any history of dyspnea at rest, ortopnea or paroxysmal nocturnal dyspnea but had dyspnea during ordinary daily activities. On physical examination there were no pulmonary rales and the hemodynamic status including heart rate, blood pressure, etc. were found to be normal during NSR. On follow-up, near- syncope bouts due to alternating attacks of SVT and Mobitz Type 2 block continued persistently. Transthoracic echocardiography (TTE) demonstrated a large perimembranous ventricular septal defect with a hypoplastic right ventricle and tricuspid atresia (Fig. 3). Left ventricular systolic function on TTE was not impaired noticeably. Left ventricular ejection fraction (LVEF) was within the lower limit of the normal value (LVEF = 50%). There was only a mild regurgitation of the atrioventricular valve. Coronary angiography was found to be normal. Electrophysiological study revealed induction of SVT with concomitant impaired atrioventricular node functions (prolonged H-V interval and emergence of 2. degree atrioventricular block). Due to the severity of symptoms, epicardial pacemaker implantation was performed. After implantation, a calcium channel blocker (verapamil) per orally was initiated for the prevention of tachyarrhythmias. Ablation of the arrhythmogenic focus was planned as a last resort in case of failure to anti-arrhythmic medication. During a week long follow-up period, frequent arrhythmias leading to severe symptoms vanished completely, and the patient was discharged with recommendation of close follow-up.

Discussion

The Fontan operation has been used as a palliative operation for pulmonary atresia, tricuspid atresia, double inlet ventricle and complex single ventricle [1]. The operation typically redirects the systemic venous return directly to the pulmonary arteries. The classical older variant (atriopulmonary anastomosis) has generally been replaced by newer modifications (lateral tunnel, external conduit) which diminish right atrial distention [1]. Atriopulmonary anastomosis generally causes remodelling, distention and dilation of the right atrium (RA) due to chronic stretching secondary to persistent pressure overloading [1] in the long term, leading to atrial conduction heterogeneity and delay. Additionally, scar tissue around the anastomosis may create a focus of re-entry. Some of the factors described above may initiate a group of atrial arrhythmias in some of Fontan patients [2, 3]. Our patient suffered initial attacks of syncope due to SVT 21 years after the Fontan operation. This interval was considerably greater than in previously reported cases. The median time to initial arrhythmia is 7 years in these patients [4]. Ablation of the arrhythmogenic focus has been successfully performed in some Fontan cases. In this case, the ablation procedure was deferred due to complete symptomatic amelioration and suppression of the tachyarrhythmias with the antiarrhythmic medication on follow-up.

An interesting point in our patient was the delayed occurrence of symptomatic heart block (21 years later) in an alternating manner together with supraventricular tachycardia. Atrioventricular node dysfunction may occur after congenital operations as a result of trauma, scar tissue or compromise of nodal blood supply. Node injuries may be reversible or irreversible and generally emerge in the early postoperative period. The mean time to pacemaker implantation after these operations was 21 days in a study [5].

In conclusion, the case presented here showed an unusually delayed form of syncopal attack due to SVT with alternating attacks of heart block. This is

a rare case demonstrating the presence of ongoing arrhythmic syncope risk throughout a patient's lifetime after operation for congenital cardiac diseases.

Acknowledgements

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