

Recovery of atrial transport function after a maze procedure for atrial fibrillation in conversion of a failing Fontan circulation

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Surgical ablation for atrial arrhythmias at conversion of atriopulmonary or ventriculopulmonary Fontan to a total cavopulmonary connection is feasible with recovery of both sinus rhythm and atrial transport function. Recovery of the patient's physical condition may take up to a year. (*Neth Heart J* 2008;16:170-2.)

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Due to atrial dilatation, atrial arrhythmias are frequent after Fontan surgery for single ventricle physiology.^{1,2} These arrhythmias can easily evolve into life-threatening events.¹ In this regard the lateral tunnel Fontan has been introduced, and Fontan constructions with an atriopulmonary or atrioventricular connection have been modified into a Fontan with a lateral tunnel or with an extracardiac conduit.³⁻⁵ These operations include surgical ablation of the atrium.^{5,6} Success is being reported with regard to recovery of heart rhythm, but the question remains whether or not the atrial transport function recovers.

Case

A male patient with the diagnosis of tricuspid atresia with atrial and ventricular septal defect (figure 1A) was stable with regard to transcutaneous oxygen saturation and physical condition at young age. At the age of 14 years a Fontan operation was carried out with closure of the atrial and ventricular septal defect and connection of the right atrium to the right ventricle with a bioprosthetic conduit (figure 1B). When he was 30, a conduit replacement by a pulmonary allograft extended with a vascular prosthesis was indicated because of conduit obstruction. At the age of 37, he experienced a decrease in exercise tolerance, resulting in New York Heart Association (NYHA) functional class 3-4. Echocardiography showed a diminished left ventricular function. The right atrium was extensively enlarged, the diameter of the inferior caval vein being 34 mm. Antegrade velocity over the conduit was 1.2 m/sec, retrograde flow 1.0 m/sec, indicating low flow with obstruction and regurgitation. Paroxysmal atrial fibrillation was diagnosed. Rhythm analysis showed accelerated atrial rhythm and atrial flutter, infrequent nonsustained ventricular tachycardia and premature ventricular beats. In this situation an acute admission for right atrial thrombosis was necessary, which was successfully treated with anticoagulant therapy.

Urgent surgery was performed. At operation, extracorporeal circulation was installed with bicaval venous cannulation. The atrial septum was resected through the right atrium. Surgical ablation consisted of a Cox III cryoablation maze procedure with resection of the left auricle and removal of all large trabeculation from the right atrium. The pulmonary valve was closed with detachment of the confluent pulmonary artery. The superior caval vein was transected, the distal end directly anastomosed to the upper side of the right pulmonary artery and the cardiac end to the caudal side of the right pulmonary artery. Because of dense adhesions of the right lung to the right atrium, an intra-atrial lateral tunnel was constructed with a polytetrafluoroethylene prosthesis. The conduit was not removed but the remnants of valve cusps were excised to allow drainage from the small right ventricular cavity

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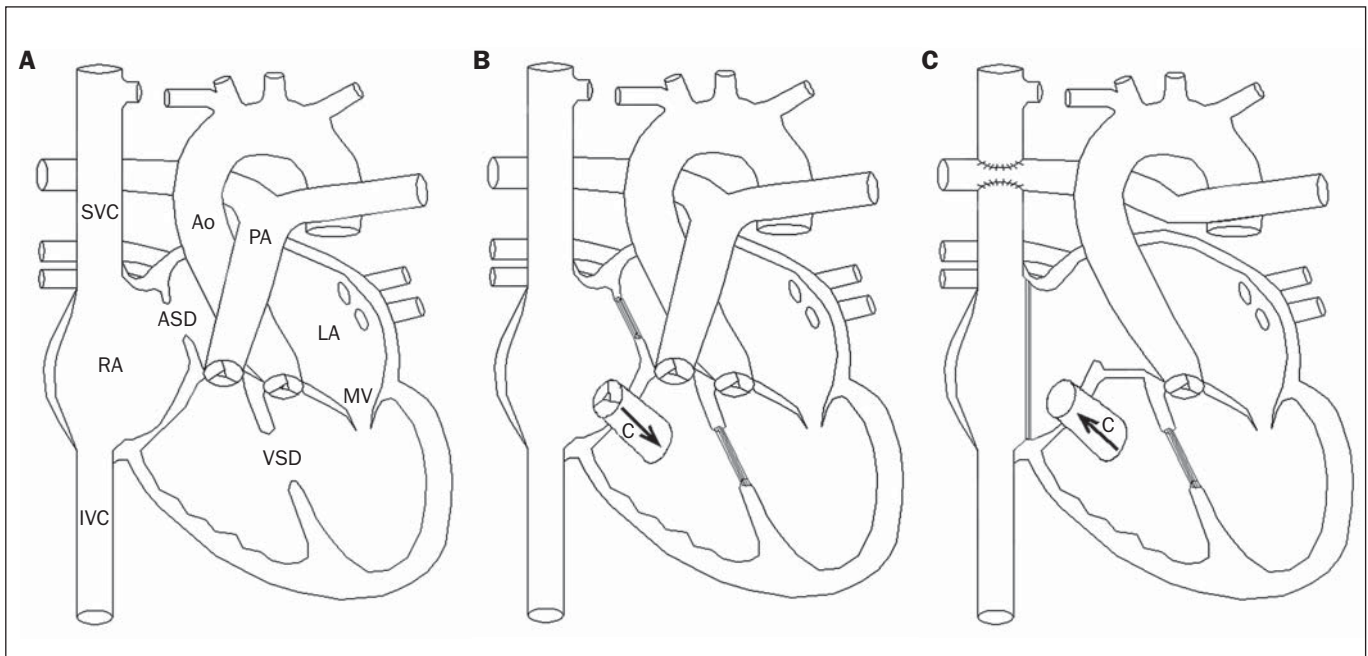


Figure 1. A) Schematic drawing of the original anatomy of tricuspid atresia with ventricular septal defect. SVC=superior caval vein, IVC=inferior caval vein, RA=right atrium, ASD=atrial septal defect, LA=left atrium, MV=mitral valve, VSD=ventricular septal defect, Ao=aorta, PA=pulmonary artery. B) Drawing of the first reconstruction. Closure of the ventricular septal defect and atrial septal defect. Valved conduit (c) interposition from the right atrium to the right ventricle. C) Drawing of the last reconstruction. Resection of the atrial septum. Removal of valve remnants in the conduit (c) to allow drainage of blood from the right ventricular cavity to the common atrium. Intra-atrial lateral tunnel as part of the total cavopulmonary connection. Closure of the pulmonary valve and disconnection of the pulmonary artery from the heart. The maze procedure and the right atrial reduction are not indicated in the drawing.

to the systemic atrium. The right atrial cavity was reduced by partly resecting the right atrial wall before closing the free edge on top of the lateral tunnel (figure 1C).

Surgery was successfully completed in sinus rhythm. The postoperative course was complicated by low cardiac output with pericardial effusion treated with a rethoracotomy to remove clots on day 2 and temporary renal failure treated with continuous venovenous haemofiltration for four days. The patient was weaned from mechanical ventilation on day 5. On day 7 an episode of atrial fibrillation was converted to sinus rhythm, again with amiodarone treatment. On day 19 the patient was discharged home.

On outpatient check-up six weeks after surgery, echocardiography showed not only persistent sinus rhythm, but also recovery of atrial transport function (figure 2). Complete recovery took about a year. The patient resumed his professional and physical activities completely. No arrhythmias recurred with digoxin and amiodarone therapy.

Discussion

In patients with single ventricle physiology, the only therapeutic option is surgical palliation by a Fontan procedure. Originally, Fontan described his operation for palliation of tricuspid atresia as a single-stage

atriopulmonary (APC) or atrioventricular (AVC) connection.³ Although a Fontan circulation with an APC or AVC may be initially successful, many patients develop complications during long-term follow-up. The most common complication in these patients is

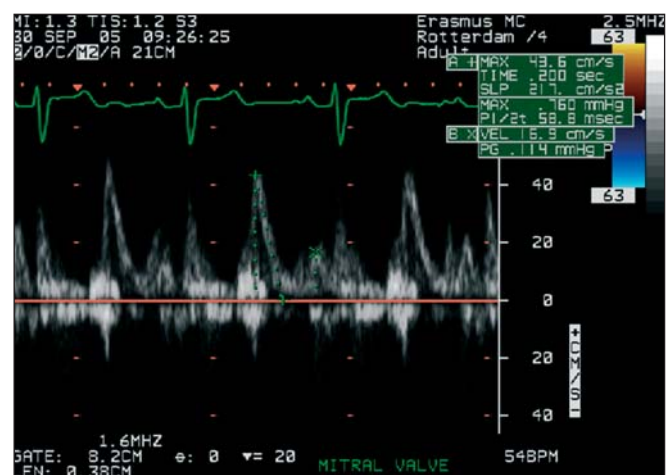


Figure 2. Echocardiographic still frame of a Doppler tracing of the mitral valve. The ECG shows sinus rhythm. The flow signal shows an E wave after the QRS complex indicating spontaneous inflow and an A wave at the electrocardiographic P wave demonstrating flow due to active transport.

progressive right atrial dilatation and consequently atrial arrhythmias, resulting in a loss of atrial transport function and a further decrease in cardiac output. In an attempt to overcome this problem, the total cavopulmonary connection (TCPC) was introduced as initial Fontan circulation.⁴

Recently the TCPC, either with a lateral intra-atrial tunnel or with an extracardiac conduit, is also being applied as a conversion for the failing APC or AVC.⁵ At conversion the atrial arrhythmias are also treated by excision of the greater part of the dilated right atrial free wall and by intraoperative ablation of potential circuits of atrial arrhythmias. Intraoperative ablation consists of a right-sided maze operation for atrial flutter or the Cox-III maze operation for atrial fibrillation.⁶ In addition, some authors propagate completion of surgery with an epicardial pacemaker system for further management of arrhythmias.⁶ The rationale is that transvenous access to the heart is no longer possible and that frequently conversion concerns a repeat sternotomy.

Mortality following these conversions of the Fontan circulation to TCPC depends heavily on patient selection and is reported to vary from 0 to 20%.⁶ After conversion to a TCPC most patients show an

improvement in NYHA class.⁶ This improvement may, however, take up to a year after surgery to become evident. Without antiarrhythmia surgery the recurrence rate of atrial arrhythmias is reported to be up to 76%.⁶ Moreover, as shown in the case presented here, recovery of atrial transport function can be accomplished. ■

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