Mustard procedure in atrial situs inversus: three-dimensional ablation of intra-atrial reentry tachycardia on a surgically created inversus cavo-tricuspid isthmus

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It has been demonstrated that the Biosense Carto™ system can improve the success rate of ablation in case of an intra-atrial reentry tachycardia in patients submitted to the Mustard repair. This system was used to map an intra-atrial reentry tachycardia in a young patient who had been submitted to the Mustard procedure for atrial situs inversus. A line of block was created connecting the right sided tricuspid valve to the left sided inferior vena cava. This terminated the arrhythmia and prevented its re-initiation. This case confirmed the notion that the cavo-tricuspid isthmus is often critical to the maintenance of an intra-atrial reentry tachycardia after the Mustard procedure even if its location is in the inversus side.

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Introduction

Intra-atrial reentry tachycardia is a severe complication of the Mustard repair[1]. Results of catheter ablative therapy, though not extremely successful, are encouraging. Recent studies suggest that a critical isthmus exists between the baffle and the ostium of the coronary sinus or between the baffle and the tricuspid annulus[2,3], practically in a location similar to that of the isthmus found in normal heart with atrial flutter.

Recently, some authors have demonstrated that the Biosense Carto™ electroanatomic mapping system is very useful in the mapping of an intra-atrial reentry tachycardia and in guiding catheter ablation[4,5].

In 1990 a combined Mustard and Rastelli operation was described as an alternative approach to associated anomalies in congenitally corrected transposition of the great arteries with ventricular septal defect and pulmonary stenosis[6]. More recently, the same combined procedure was reported in 3 patients with the same complex disease but in atrial situs inversus[7].

We describe a case of an intra-atrial reentry tachycardia which developed as a late complication of this procedure. The Carto™ system was the only system used to map and ablate the circuit responsible for the tachyarrhythmia. In particular, the existing surgically created "inversus isthmus" between the left sided inferior vena cava and the right sided tricuspid annulus was ablated by means of a linear lesion.

Case report

An atrial situs inversus, corrected transposition of the great arteries, an interventricular septal defect and pulmonary valve stenosis were diagnosed in a 15-day-old child (C.F.). At the age of 9 years, he underwent the Mustard and Rastelli procedure (a patch of teflon used to close the interventricular septal defect, a patch of Gore-tex used for intra-atrial rerouting after the demolition of the atrial septum, a patch-tunnel of filamentous Dacron fabric used for atrial baffle and a 24 mm composite valved homograft employed to establish continuity between the right ventricle and the pulmonary artery) (Fig. 1). After a several-month history of brief episodes of palpitations, at the age of 13 years, the patient was admitted to our hospital for palpitations and asthenia. Standard electrocardiography showed an atrial tachycardia with negative atrial deflections in leads II, III, and aVF (Fig. 2). After a successful cardioversion by means of transesophageal

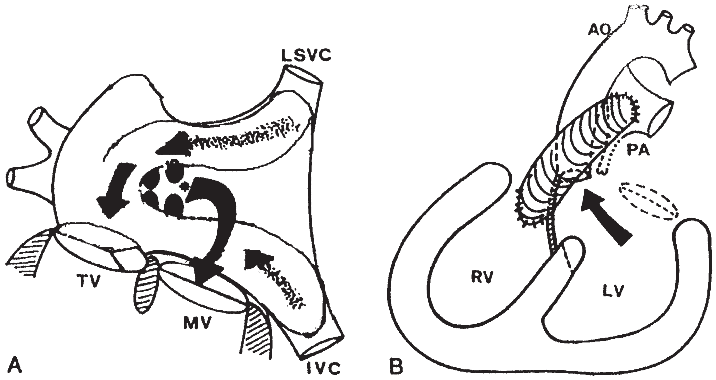

Figure 1. *The figure shows a schematic two-dimensional representation of the Mustard-Rastelli operation and the direction of the blood flow in the heart chambers.* A: *Mustard operation performed through the left sided right atrium.* B: *Rastelli operation including patch closure of the ventricular septal defect through the right sided ventricle and placement of a valved conduit between the right ventricle and the main pulmonary artery. AO = aorta; IVC = inferior vena cava; LSVC = left superior vena cava; LV = left ventricle; MV = mitral valve; PA = pulmonary artery; RV = right ventricle; TV = tricuspid valve. * = pulmonary veins.*

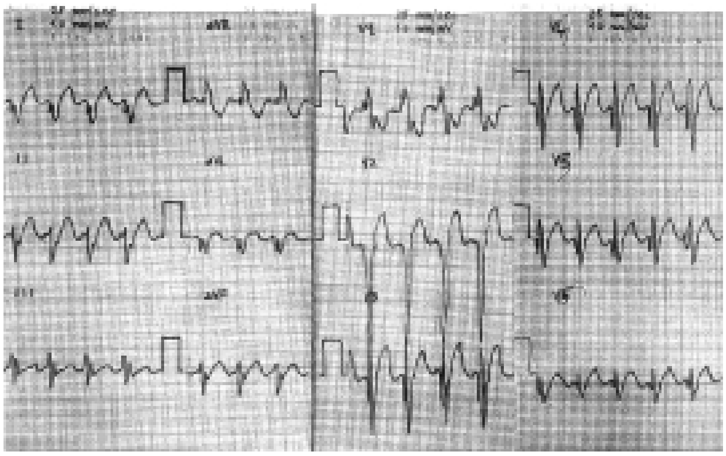

Figure 2. *Standard 12-lead electrocardiogram of the intra-atrial reentry tachycardia.*

overdrive pacing, drug therapy with digoxin and amiodarone was started and the patient had no recurrences for about 5 years.

At the age of 18 years, he complained of sudden-onset palpitations. A recurrence of the same atrial tachycardia was diagnosed and the patient was again submitted to an electric cardioversion. Following complete pharmacological wash-out, we decided to submit the patient to hemodynamic evaluation and mapping and ablation of the atrial tachycardia. Written informed consent was obtained for the procedure. The patient was evaluated during general anesthesia, induced with sevofluorane and propofol and maintained with sevofluorane or isofluorane.

Hemodynamic evaluation and angiography (Fig. 3) revealed the following residual anomalies: a moderate stenosis in the superior caval conduit of the Mustard repair with a mean gradient of 15 mmHg, a mild stenosis of the right ventricular-pulmonary artery conduit with a gradient of 25 mmHg and, finally, two small residual interventricular defects (patch detachment).

A 5F quadripolar Josephson catheter and a 7F Navistar mapping and ablation catheter (Biosense-Webster, Haifa, Israel) were introduced through the right femoral vein up to the systemic venous atrium. The intra-atrial reentry tachycardia with an atrial cycle length of 260 ms and a 2:1 atrioventricular conduction was induced and then mapped using the Carto^{TM} system. The distal bipolar atrial signal from the quadripolar catheter, placed laterally in the systemic atrium, was used as timing reference for the activation mapping. A window of interest equal to the cycle length of the tachycardia was established around the timing reference signal. Before the procedure, the navigation catheter was placed outside the patient's chest and deflected in all directions within the magnetic fields in order to coordinate the tip deflection color. Having established the fundamental anatomical and electrophysiological landmarks such as the two caval veins and tricuspid annulus, a three-dimensional electro-anatomic map of the atria was reconstructed from the multitude of the endocardial sites that were sequentially mapped.

The intra-atrial reentry tachycardia showed an anticlockwise movement around the tricuspid valve annulus with an intact and homogeneous conduction between the left sided inferior vena cava and the tricuspid annulus via the isthmus (Fig. 4). Therefore, by means of 25 radiofrequency applications (preselected temperature 70°C, duration 60 s) (Fig. 5), a linear lesion connecting these two structures was made and the tachycardia was successfully interrupted (Fig. 6). Unfortunately, the particular anatomy made the identification

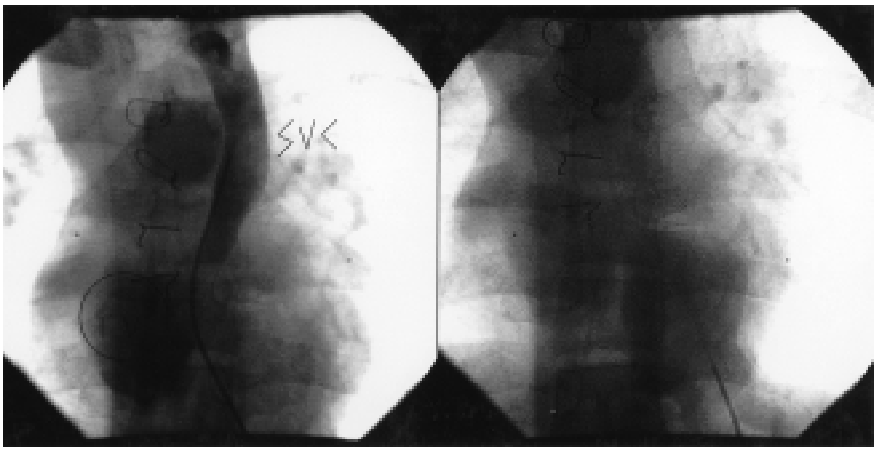

Figure 3. *The angiogram (antero-posterior view) of the new systemic atrium depicted by the Swan-Ganz catheter through the left sided inferior and superior vena cava (SVC). TV = tricuspid valve.*

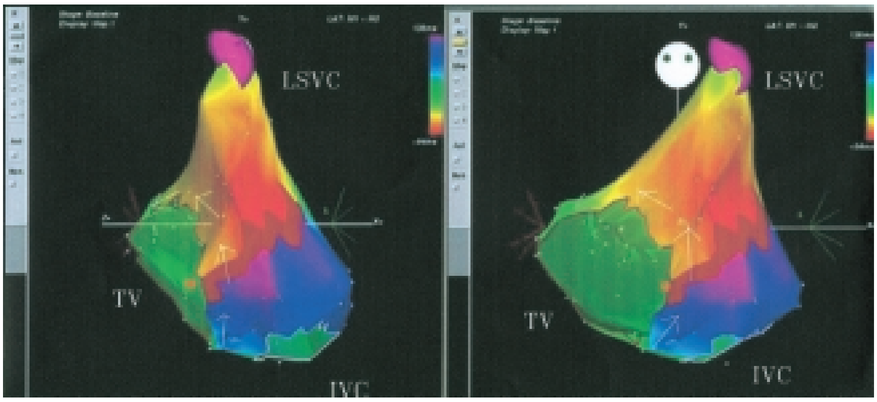

Figure 4. *Activation map (left anterior oblique and antero-posterior views) of the new systemic atrium during intra-atrial reentry tachycardia before ablation. The red and purple areas respectively denote the sites of early and delayed activation. The map shows a new systemic atrial activation propagating around the tricuspid valve annulus in an anticlockwise direction. Abbreviations as in figure 1.*

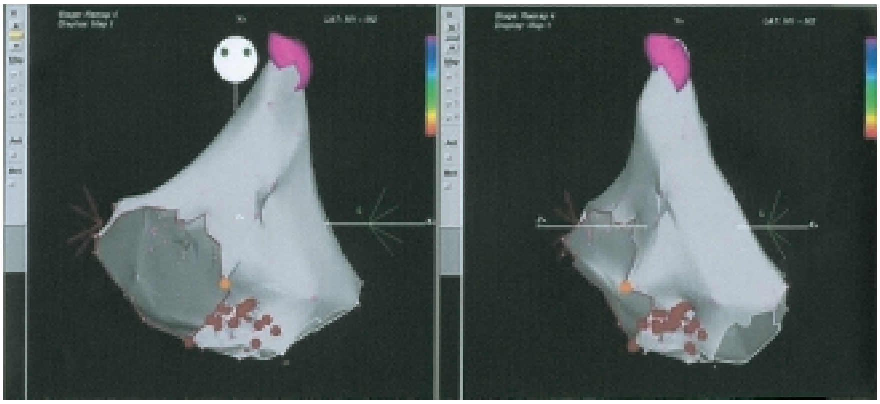

Figure 5. *Red tags represent the radiofrequency applications forming the linear lesion which interrupted the intra-atrial reentry tachycardia on the "inversus isthmus".*

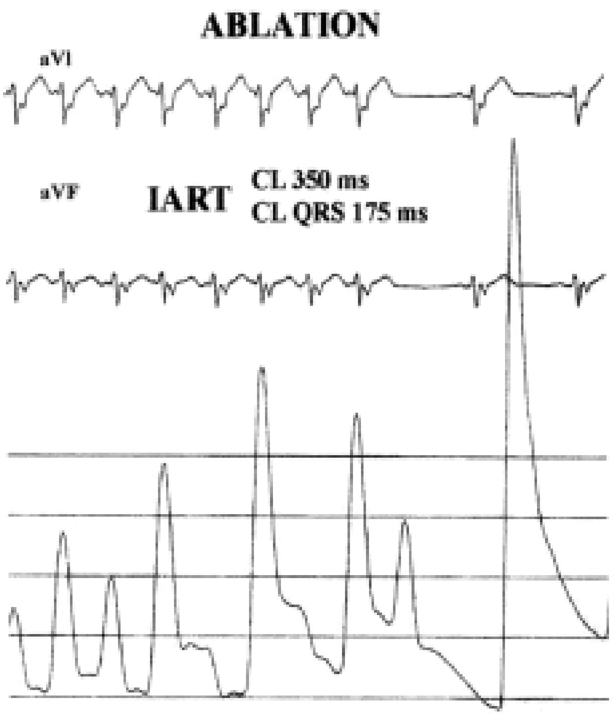

Figure 6. *Tachycardia interruption during radiofrequency ablation. CL = cycle length; IART = intra-atrial reentry tachycardia.*

and evaluation of a bidirectional block along the linear lesion difficult and for this reason it was not performed.

However, during the next hour, tachycardia was no longer inducible neither in basal conditions nor during isoproterenol intravenous infusion (0.01-0.04 μg/kg/min).

The total time of the procedure was 435 min and the total time of fluoroscopy was 60 min.

After 24 hours the patient was discharged in good clinical conditions. No antiarrhythmic drugs were prescribed. The intra-atrial reentry tachycardia never recurred during the 14-month follow-up.

Discussion

Congenitally corrected transposition of the great arteries in situs inversus is the mirror image variant of the more frequent type in situs solitus. It is almost uniformly associated with a ventricular septal defect and pulmonary outflow tract obstruction. Physiologic repair of this anomaly, leaving the right ventricle in support of the systemic circulation, has been accomplished by means of different techniques of ventricular septal defect closure combined with relief, or bypass, of the pulmonary outflow tract obstruction. After the report by Ilbawi et al.[6] of the successful cardiac redirection of the venous and arterial flows in patients with congenitally corrected transposition of the great arteries in situs solitus, our group[7] reported, in 1992, 3 cases of congenitally corrected transposition of the great arteries in situs inversus in which the same principle of anatomic repair was adopted. In the discussion of that report the following statement was made: "The true liability of this technique, compared with those currently used for physiologic repair, lies in the potential introduction of the well known problems associated with extensive intra-atrial baffle procedures (e.g., arrhythmias and pulmonary venous obstruction)". And, in fact, confirming what was declared, after 10 years in this case report we describe the clinical history, the evaluation and treatment of an intra-atrial reentry tachycardia occurred in one of these 3 cases during the postoperative follow-up.

Our case seems to confirm previous experiences of other authors describing the importance of the Carto™ system in helping to understand and treat the substrate of the intra-atrial reentry tachycardia after correction or palliation of complex congenital heart diseases[4,5,8,9]. Moreover, in this particular case, Carto™ was the only system utilized to evaluate and to eliminate the tachycardia.

It is well known that the Carto™ system also allows an important reduction in radiological exposure and this was confirmed even in our case in which the cardiac anatomy was very complex. In fact, the fluoroscopy time, inclusive of cardiac catheterization, was only 60 min which is about one seventh of the total procedure duration.

Recently the cavo-tricuspid isthmus has been identified as a critical area for the ablation of an intra-atrial reentry tachycardia in patients who had previously been submitted to the Mustard procedure[2-5,8] and more recently, Love et al.[8], using the Carto™ system, have demonstrated that the tricuspid annulus is the more frequent "central obstacle" of the postoperative intra-atrial reentry tachycardia in case of biventricular correction of complex congenital heart diseases.

In our particular case the tricuspid annulus was again the central obstacle of the mapped intra-atrial reentry tachycardia, but interestingly the cavo-tricuspid isthmus, surgically created by reconstructing the new systemic venous atrium, was not in the usual location, but on the inversus side. In fact, it was located between the tricuspid valve, in the correct anatomical position, and the orifice of the inferior vena cava, in a left position.

The particular anatomy and surgical reconstruction permitted complete mapping of the area even though the access of the mapping catheter was possible only in the new systemic atrium. A radiofrequency linear lesion transecting this "inversus cavo-tricuspid isthmus" resulted in the interruption and disappearance of the intra-atrial reentry tachycardia.

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