

## Atrial Flutter In A Tetralogy Of Fallot Operated Patient: Importance Of A Rapid And Curative Treatment

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### Abstract

A 51 male, affected by Tetralogy of Fallot, underwent a left Blalock-Taussig anastomosis at the age of two years and an aorto - right pulmonary artery tube graft when 8 years old. Complete surgical correction was performed at age 21 with closure of the ventricular septal defect and a large patch over the right outflow tract, shunts were discontinued. Then it was well up to 51 years old when he began to suffer shortness of breath with minimal exertion. with ECG evidence of supraventricular tachycardia. Suggestive signs of a typical atrial flutter led to early electrophysiological assesment and successful cavo-tricuspid isthmus ablation was successfully performed. Echocardiographic and magnetic resonance imaging and ergospirometry provided complete informations on anatomic and hemodynamic conditions but no other interventional procedure was necessary.

### Introduction

Tetralogy of Fallot (ToF) is the result of anterior malalignment of the conal septum during embryological development. It is the most common cyanotic congenital heart defect, with an incidence rate of 32.6 per 100,000 live births.<sup>1</sup> Complete surgical repair has successfully been performed since 19542, but long-term outcome may be complicated by atrial and ventricular arrhythmias.<sup>3-6</sup> Patients over 50 years are rare and a very long term follow up has yet to be written. Conduction and rhythm abnormalities are associated with morbidity and increased mortality.<sup>5</sup> Atrial tachyarrhythmias (AT) such as atrial flutter or fibrillation are seen with increasing frequency beginning from the third decade of life.<sup>6</sup> We hereby describe a rare case of very long term follow up complicated by atrial flutter successfully treated with radiofrequency ablation.

### Case Presentation

A 51-year-old male affected by an operated Tetralogy of Fallot was admitted to our Cardiological department for dyspnea on effort. When two year-old he underwent a Blalock-Taussig (BT) anastomosis as a palliation. He did relatively well until the age of 8 when he started having progressive symptoms with cyanotic spells for which reason, in right thoracotomy, a second shunt between ascending aorta and right pulmonary artery with a 7 mm tube graft was performed after which he did relatively well. At age 21 he manifested signs of increased cyanosis, marked limitation of activity and headache. He underwent cardiac catheterization whose findings

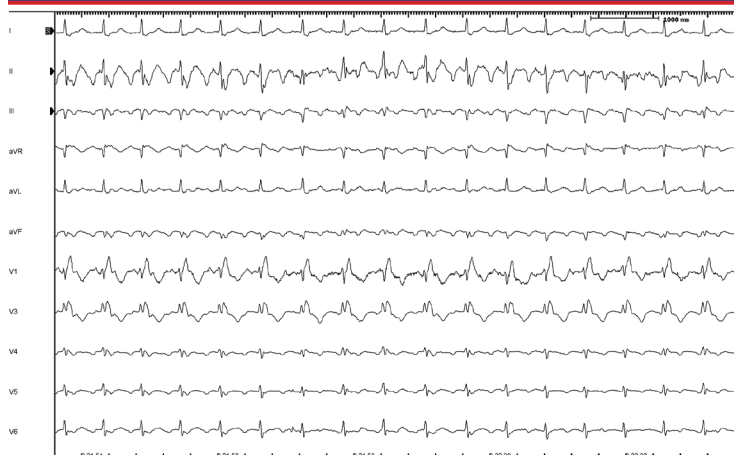
were consistent with severe pulmonary stenosis, closed left BT shunt and poorly functioning aorto pulmonary shunt. Surgical correction was performed at the Texas Heart Institute, Houston, with closure of the ventricular septal defect and a large patch over the right outflow tract, which was found patent at surgery, the aorto pulmonary shunt was discontinued. No arrhythmias were present in the perioperative period.

The patient did relatively well until 51 years old when he developed progressively worsening dyspnea and was admitted in our Cardiological Department with the diagnosis supraventricular arrhythmia. The transthoracic echocardiogram showed a severely dilated right ventricle with the proximal outflow tract end-diastolic diameter 64 mm from the parasternal long axis view, the basal end-diastolic right ventricle diameter from the 4 chamber view was 63 mm, the left atrial are was 30 cmq and the right atrial area 34 cmq. The right ventricular free wall thickness was increased (6 mm) and the contractility moderately depressed with a fractional area change of 30% and a TAPSE (Tricuspid Annulus Systolic Excursion) of 16 mm. Tricuspid annulus S wave velocity was 10 cm/sec. The right ventricular outflow tract (RVOT) was not obstructed, the pulmonary valve insufficiency was moderate. A mild tricuspid insufficiency was present and the estimated ventricular systolic pressure was 38 mmhg. No residual ventricular patch shunt was present. The left ventricle ejection fraction was 54%. A transesophageal echocardiogram excluded auricular thrombosis.

12 lead ECG showed regular atrial arrhythmia at 105 bpm with a right bundle branch QRS morphology, regular atrial activation with inverted saw-tooth F-wave pattern in inferior ECG leads II, III, and aVF with low amplitude biphasic F waves in leads I and aVL, and upright F wave in precordial lead V1 and inverted F wave in lead V6 (Figure 1). These features suggested an isthmus dependent

**Disclosures:**  
None.

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#### Surface 12 lead electrocardiogram.

Lateral, Augmented and Precordial leads shows atrial flutter with right bundle branch block morphology. F waves have saw-tooth pattern in inferior leads, flat aspect in I and aVL, and upright F pattern in V1 suggesting a typical atrial flutter.

Figure 1:

atrial flutter, therefore an electrophysiological study was performed during ongoing arrhythmia. A decapolar deflectable mapping catheter was advanced into the coronary sinus showing proximal-to-distal activation with 2:1 atrial-ventricular conduction. Measured cycle was 286 msec. Ablator Catheter (Navistar DS 8 mm Biosense Webster, Diamond Bar, CA) was introduced in the right atrium and electro-anatomic map of the chamber was reconstructed using 3D computerized activation system (CARTO system Biosense Webster, Diamond Bar, CA). The activation and propagation maps showed counterclockwise activation pattern around the tricuspid valve with the typical aspect of early-meets-late at the cavo-tricuspid isthmus (Figure 3, Video 2). Pacing from the CTI demonstrated concealed entrainment thus confirming diagnosis of typical isthmus-dependent atrial flutter.

Application of radiofrequency (RF) (50 Watt, 65°) in the isthmus area (6 o'clock) determined arrhythmia interruption after 60 sec (Figure 4). Persistency of bidirectional conduction was verified. Detailed mapping of the ablation line was repeated identifying local conduction breakthroughs. Two selective applications of RF (320 sec each one) at these points led to complete and definite interruption of isthmus conduction (Figure 5). Results persisted after 20 minutes. The procedure terminated successful without acute complications. The ecg showed a sinus rhythm with atrio-ventricular first degree block, right ventricular hypertrophy and right bundle branch block with a QRS duration 180 ms.

Once in sinus rhythm, a cardiac magnetic resonance (CMR) and an ergospirometric evaluation was performed according to guidelines<sup>7-8</sup>. CMR showed an enlarged left ventricular volume: 83 ml/mq with mildly reduced ejection fraction: 50 %; dilated aortic root: 45 x 38 mm; right ventricular volume: 133 ml/mq (z-value 3,3) with reduced ejection fraction: 37 %. The pulmonary infundibulum was dyskinetic due the presence of large patch but without significative delayed enhancement (Figure 2 Panel C). The pulmonary valve was present with moderate insufficiency and the regurgitant fraction was 31% (Video 1).

The ergospirometric study was interrupted after 13.2 minutes at the workload of 125 Watts due to dyspnea (Borg 4) but without arrhythmias. Peak O<sub>2</sub> consumption was 73 % of the maximal predicted

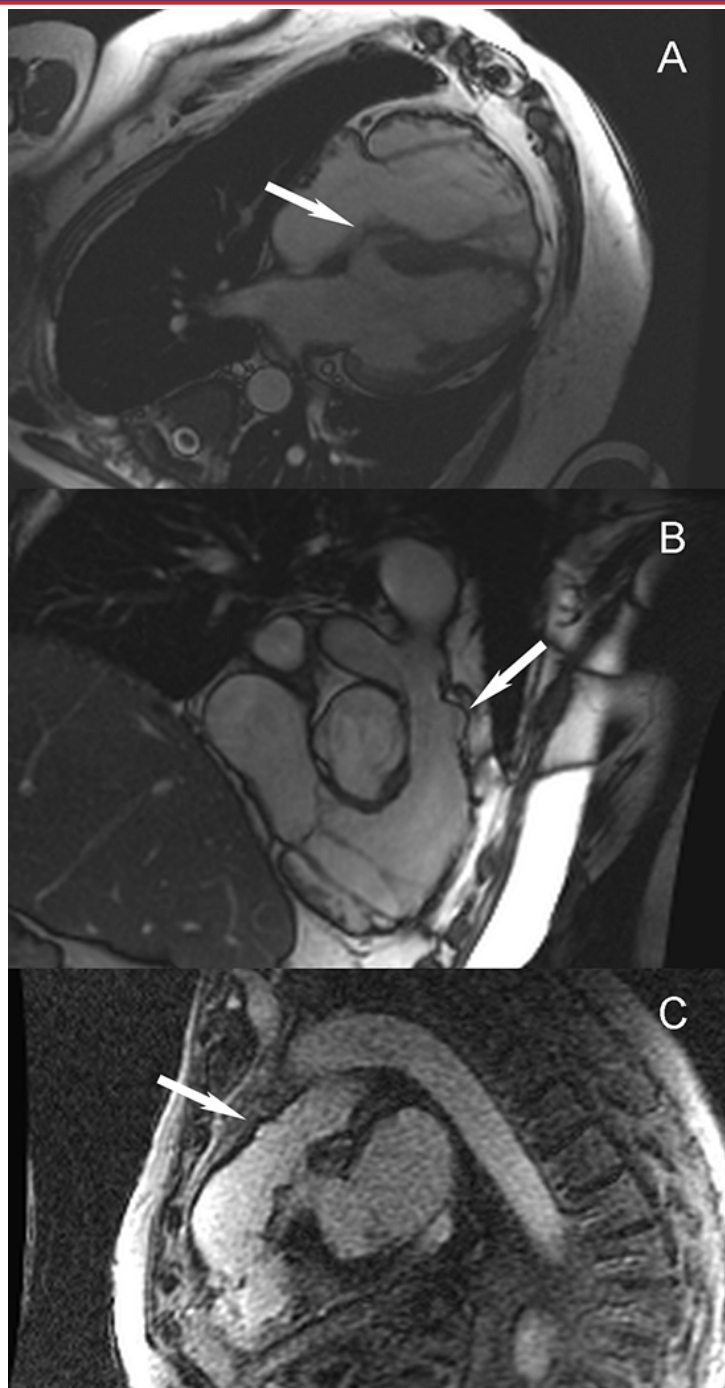


Figure 2:

#### Cardiac magnetic resonance.

Panel A: steady state free precession still image, four chamber view shows the dilated right atrium and ventricle and the interventricular patch (arrow).

Panel B: steady state free precession still image, right ventricle 2 chamber view shows the inflow and outflow tract with the aneurismatic patch (arrow).

Panel C: T1 weighted post-gadolinium sequence shows absence of significant enhancement in the right ventricular outflow tract (arrow).

value: peak O<sub>2</sub>/Kg 22.9 mL/kg/min, VE/VCO<sub>2</sub> slope.<sup>25</sup> VO<sub>2</sub>/work, slope pari a 11, Peak Respiratory Exchange Ratio (RER).<sup>1,19</sup> The anaerobic threshold was achieved at 44 % of the maximal predicted O<sub>2</sub> consumption (59 % of the peak O<sub>2</sub> consumption). No desaturation was present during the test.

After the ablation procedure the patient has been well without

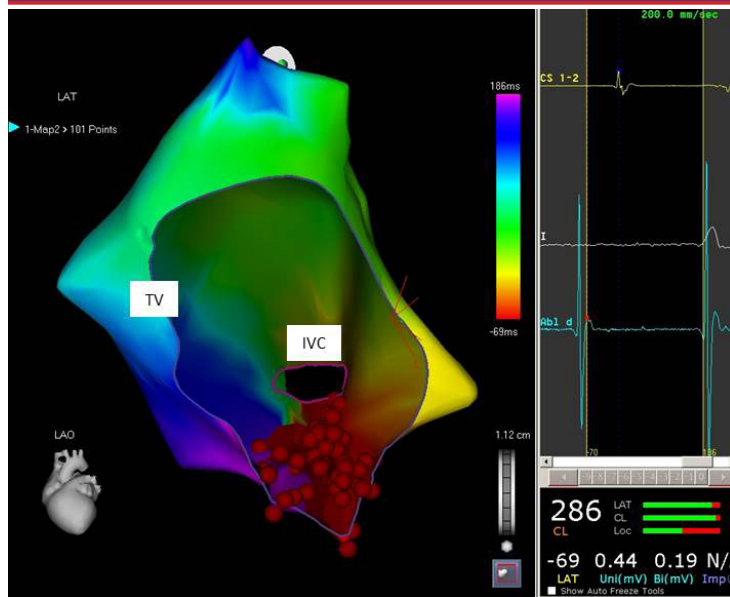


Figure 3:

Left Anterior Oblique (LAO) view of three-dimensional electro-anatomic map (CARTO, Biosense Webster, Diamond Bar, CA). During ongoing arrhythmia with atrial cycle of 286 msec the map shows a typical counterclockwise activation pattern around the tricuspid annulus which is based on a color scheme indicating activation time from orange (early) to purple (late). The two colors are closed in the region of isthmus (early-meets-late) were red dots indicate ablation line. The right side of the figure shows intracardiac recording signals from coronary sinus catheter (CS 1,2) and ablator (ABLd) and surface activation from I lead. Ablator, positioned in the isthmus region ablator displays two activation potentials respectively at the beginning and at the end of the cycle, atrial activation on the CS catheter is approximately mid-diastolic.

significant limitations to his physical activity and no arrhythmia recurrence.

## Discussion

In ToF surgical relief of the RVOT obstruction involves infundibulotomy, resection of obstructive muscle bundles and the use of a patch to enlarge the pathway from the right ventricle to the pulmonary arteries. These procedures result in scar tissue and akinetic

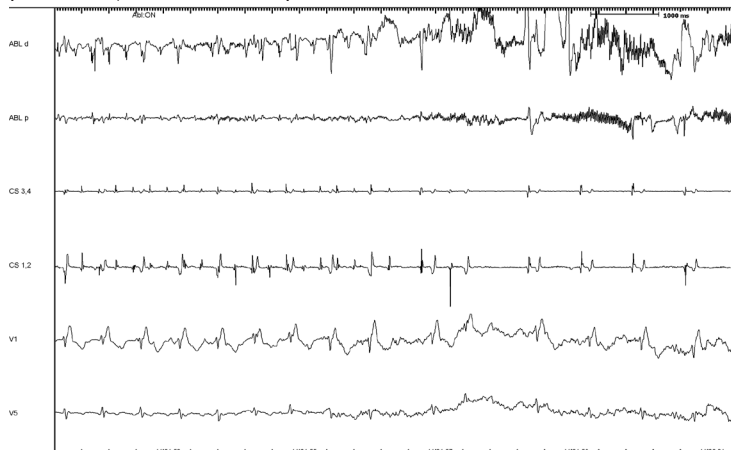


Figure 4:

Arrhythmia interruption during radiofrequency application. Endocardial electrocardiogram from the ablator (ABLd, ABLp) and coronary sinus catheter (CS 3,4, CS 1,2). Surface activation from precordial leads (V1 and V6). 12th ventricular activation is in sinus rhythm. Real time cardioversion of atrial flutter happened 60 sec after initiation of radiofrequency applications.

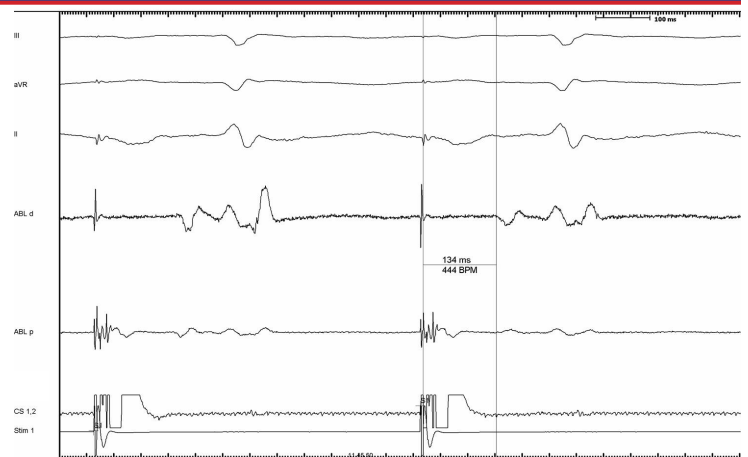


Figure 5:

Confirmation of isthmus block after radiofrequency ablation. Surface electrocardiograms from III, aVR, II endocardial electrogram from the Ablator (ABLd, ABLp) and coronary sinus (CS 1,2). Activation on the ablator catheter lateral to the ablation line happens 134 msec after pacing stimulus from coronary sinus (S1), thus confirming a conduction block. Similar results were obtained with pacing stimulus from the ablator with activation signals sampled on the coronary sinus, thus showing bilateral conduction block.

RVOT free wall, which can become aneurysmatic, and pulmonary valve insufficiency. Other abnormalities that may be found in patients with repaired ToF are branch PA stenosis, residual atrial or ventricular septal defect, tricuspid regurgitation, right ventricular dilatation and dysfunction, aortic dilatation, aortic regurgitation, and left ventricular dysfunction. Corrective surgery requires ventricular and atrial incisions, therefore incisional reentrant tachycardias may develop, but the majority of AT are right atrial macro-reentrant tachycardias, most often involving the cavo-tricuspid isthmus<sup>9</sup>. Whatever the underlying electrophysiological mechanism, AT in TOF patients have a significant impact on clinical outcome and are associated with congestive heart failure, stroke, and death.<sup>3-6</sup> Even in a such distorted anatomy as in ToF, classic ECG signs of isthmus-dependent atrial flutter can lead to perform earlier electrophysiological evaluation to treat and rapidly solve the arrhythmia, due to its high therapeutical success (98%).

## Conclusions

Development of atrial tachyarrhythmias in repaired ToF patients causes a severe worsening of the hemodynamic state and clinical outcome. This case report underlines the usefulness of an early invasive approach as effective and rapid curative treatment when the arrhythmia has typical signs of isthmus-dependent of atrial flutter which is an easy to ablate arrhythmia. After stabilization of hemodynamic compromise it highlights and the need of a complete evaluation of the post-operative anatomical and functional status.

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