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Arrhythmias And Long-Term Hemodynamic Consequences In Patient With Repaired Tetralogy Of Fallot – A Case Study

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Abstract
The number of patients who underwent total correction of Tetralogy of Fallot (ToF) in early life or infancy is still rising. According to CDC ToF is the most prevalent cyanotic congenital heart disease with incidences of 1 in 2500 births. Rhythm disturbances and haemodynamic disturbances including valves disorders, heart failure, residual defects and more are new challenges for clinicians taking care of adult patients with repaired ToF. Our goal was to describe such a patient and highlight further complications which may be encountered among those patients.

Key words: repaired tetralogy of Fallot, arrhythmia, leakage through septal patch

Introduction

ToF is a common congenital heart defect and as the number of adults with repaired ToF is growing, the new problems emerge. A regular monitoring of health among those patients is crucial for early diagnosis of problems such as irregular heart rhythm, pulmonary regurgitation, or right ventricle failure [1].

Case presentation

A 46-year-old woman with repaired ToF, implanted dual-chamber peacemaker (PM) due to complete heart block, asymptomatic atrial fibrillation (AF) lasting for two months, detected by PM and heart failure with mildly reduced ejection fraction (HFmrEF) was administered to the hospital with a feeling of heart palpitations, limited exercise tolerance and decreased well-being. Her medical history included electrical cardioversion due to atrial flutter (AFL) in 2009, moderate left to right leakage through septal patch with features of pulmonary arterial hypertension (PAH).

On physical examination the patient had a loud murmur best heard in pulmonic valve post and Erb's point, audible in all listening posts, blood pressure (BP) 132/70mmHg, regular heart rate (HR) of 70 beats per minute (bpm). She underwent transoesophageal echocardiogram which did not show any visible clot in heart cavities. After that an electrical cardioversion of AF in short anaesthesia was performed, resulting in restoring of sinus rhythm. Six month later same patient was administered with another strong heart palpitations feeling, rising in the past four days preceding administration. On physical examination she was alert and oriented with BP 113/70 and HR 70 bpm. During hospitalisation, transoesophageal echocardiogram (TEE) which excluded any clot in heart cavities, and ablation of paroxysmal atrial tachycardia in the range of right atrium and inferior isthmus of typical atrial flutter with CARTO system were performed (Figures A-C). The procedure resulted in sinus rhythm restoration. Patient was discharged without any complications. In follow-up consultations 11 days and 3 months after hospitalization no signs of any arrythmia were detected.

Discussion

About 20% of adults with repaired ToF will develop atrial arrhythmia [2-4]. Intraatrial reentrant tachycardia (IART) is second (after ventricular tachycardia) most common single arrhythmia subtype followed by AF in this population. However collectively, atrial arrhythmias are more prevalent than ventricular arrhythmias. Nevertheless, after the age of 55 years AF exceeded IART. The sharp increase in occurrence of AF develop noticeably earlier in patients with repaired ToF (<45 years of age) than in general population (>65 years of age) [5, 6]. Our patient's first diagnosed arrhythmia was AFL when she was 34 years of age.

Transthoracic Echocardiography (TTE) has been performed in our patient and showed both enlarged atria and both ventricles. Moreover, she was diagnosed with HFmrEF and her medical history includes both AF and AFL. It has been recognised that at least 25% of patients after the repair of Tof will develop left ventricle (LV) dysfunction at long-term follow-up. In this population atrial arrhythmias were associated with tricuspid regurgitation and lower right ventricle ejection fraction [7-10]. Presented patients left ventricle ejection fraction (EF) was shown to be 48% additionally, moderate tricuspid regurgitation, systolic dysfunction of right ventricle with EF equal to 38,5% and right ventricular global longitudinal strain up to -13,2% were also detected.

Retrospective study based on a group of 113 patients with repaired ToF and suffering from atrial arrhythmias, has shown that the time needed for atrial arrhythmias to resurface after cardioversion ranges from 4 to 38 months. Of 113 patients 22 underwent catheter ablation resulting in 91% success rate, without arrhythmia post procedure. It has been pointed out that AFL can be a common condition typically regarding RA, whereas AF should draw attention of clinicians because it is associated with reduced long-term survival [11, 12]. Contrary to the mentioned study, atrial arrhythmia in presented patient resurfaced 12 years after successful cardioversion which was performed in 2009.

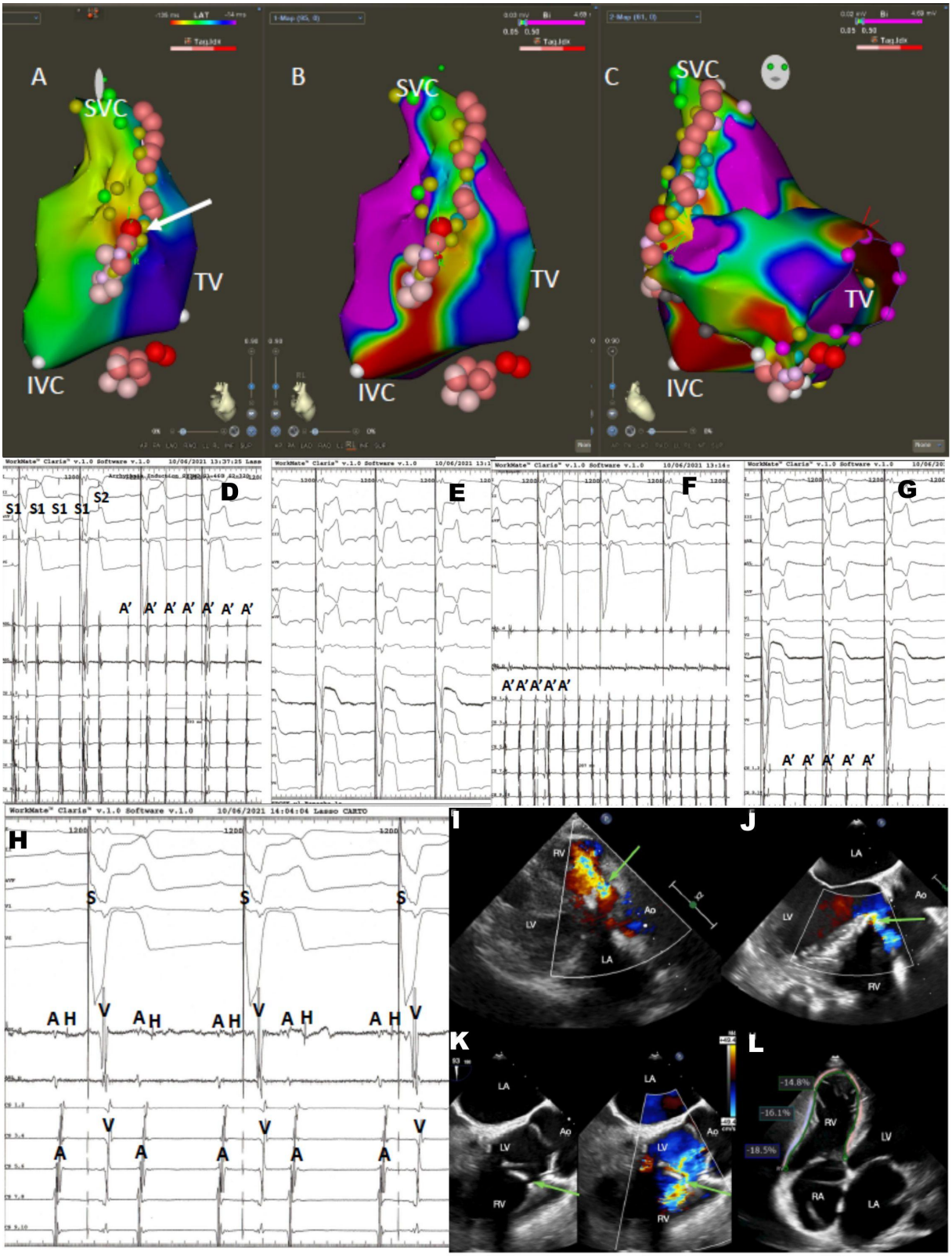
Components of atrial tachycardia (AT) in our patient were reentry loop around a scar on a lateral side of RA and the area of fast pathway within this loop. AT in patients with repaired ToF can be treated with radiofrequency ablation with acute success rate of 98%. The number of heart surgeries, age and the occurrence of AF are the predictors of recurrence. Patients at the age of 55 or less during the procedure and LA area less or equal 30 cm^2 are the factors of recurrence-free survival. It has been observed that the presence of LA tachyarrhythmia or AF is associated with inferior rhythm status at follow-up [13, 14]. In our patient ablation was performed when she was 46 yeas of age, the procedure was successful with no symptoms of atrial arrhythmia to this day.

Data from four European centres versed in tachyarrhythmias ablation in patients with congenital heart defect has shown the acute success rate of 98%. Thus, subsequent procedure was required in 16% of patients in long-term outcome. Moreover, 51 out of 52 AT present in ToF patients and studied in this paper, originated from scars and incisions created during complete repair. It has been suggested that extending the atriotomy incision towards the inferior vena cava during cardiac surgery in addition with surgical ablation can be efficacious in preventing AT in further perspective [13, 15, 16].

PR has been identified as one of the most frequent substrates of sustained ventricular tachycardia (SVT), which is equated with sudden death that occurs in up to 6% patients late after repair. Evidence has been demonstrated

that pulmonary valve replacement (PVR), in patients with repaired ToF and diagnosed with PR or right ventricular outflow tract obstruction (RVOTO), stabilizes QRS duration and in association with intraoperative ablation, reduces the occurrence of pre-existent atrial and ventricular tachyarrhythmia [17-19]. PVR is IC recommendation of European Society of cardiology in symptomatic patients with serious pulmonary regurgitation or moderate RVOTO. In patients with no native outflow tract and if anatomically feasible, transcatheter pulmonary valve implantation (TPVI) should be performed [2, 20].

Residual ventricular septal leakage has been shown as the most common indication for reoperation, responsible for almost half of the cases and followed by PVR. However, major part of patients who were responsible for this conclusion, have undergone corrective surgery in the early era of ToF surgical repair [21, 22]. It has been shown that residual VSD smaller than 2mm are likely to close itself. The ones greater than 2mm most presumably will not close spontaneously, but they are mostly hemodynamically and clinically insignificant. After VSD closure endocarditis prophylaxis is indicated and when residual shunt is greater than 2mm it might last thorough one's life [23-25]. Residual left to right leakage through septal patch in presented patient was visualised in the upper part of ventricular patch (visualised on figures I-L). Its maximal pressure gradient was ascertained to be 86mmHg which was classified as negligible, asymptomatic and any subsequent intervention was abandoned however indication for infective endocarditis antibiotic prevention exists.



Figures legend

A. Local activation time map during atrial tachycardia. The right lateral projection. The earliest activation is red while the latest is violet. The arrow shows „early meets late" area critical point for reentrant loop.
B. Potential map of the right atrium. The right lateral projection. Red – scar, violet – healthy muscle, other colors – low voltage area.
C. Fusion map from B picture and map of the inferior part of right atrium performer for cavo-tricuspid line. The RAO projection. Small pink points – fragmented potentials, small blue points – double potentials

area, small yellow points – pacing sites without phrenic nerve stimulation, small green points – phrenic nerve stimulation. Bigger pink and red points – ablation points (color depends on „ablation index" value: borders 350 and 550. IVC – inferior vena cava, SVC – superior vena cava, TV – tricuspid valve).

D. Atrial tachycardia (CL 393ms) induction with programmed pacing (S1-460ms, S2 320ms) from the coronary sinus. I, II, aVF, V1, V6 – classical ECG leads, A' – local atrial activation during atrial tachycardia, Abl – electrogram from mapping catheter, CS – electrograms from the catheter in the coronary sinus.
E. Typical atrial flutter with very slow conduction in cavotricuspid isthmus (with isoelectric line between consecutive atrial activation meets electrocardiografic criteria for atrial tachycardia).
F. Typical atrial flutter with very slow conduction in cavotricuspid isthmus. I, II, aVF, V1, V6 – classical ECG leads, A' – local atrial activation during atrial flutter, Abl – electrogram from mapping catheter, CS – electrograms from the catheter in the coronary sinus,
G. Atrial tachycardia (CL 393ms). I-III, aVR, aVL,aVF, V1-V6 – classical ECG leads, A' – local atrial activation during atrial tachycardia, CS – electrograms from the catheter in the coronary sinus,
H. Permanent ventricular pacing because of distal the third degree atrio-ventricular block. I, II, aVF, V1, V6 – classical ECG leads, A – local atrial electrogram, Abl – electrogram from mapping catheter localized in His bundle region, CS – electrograms from the catheter in the coronary sinus, H – His bundle electrogram, S – stimulus from the ventricular pacemaker, V – local ventricular electrogram
I. 2D TTE, Color Doppler
J. 2D TEE, Color Doppler
K. 2D TEE, Color compare
L. 2D TTE, right ventricle strain Arrows show ventricular septal defect Ao- ascending aorta; LA- left atrium; LV- left ventricle, RA- right atrium, RV- right ventricle

Conclusion

Adult patients who underwent ToF correction early in life are a growing and challenging population. This group is at risk of atrial and ventricular rhythm disturbances, haemodynamic impairment, heart failure and other conditions such as residuals defects. While taking care of such a patient, cardiovascular examination, and additional studies e.g., TTE, ECG monitoring, can give the clinician an overview of the patient's medical conditions and reveal any signs of potential abnormalities.

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