

## COMBINED INTERVENTIONAL TREATMENT OF ADULT PATIENTS WITH REPAIRED TETRALOGY OF FALLOT: FROM ELECTROPHYSIOLOGICAL STUDY TO PULMONARY VALVE REPLACEMENT

L. Ridolfi<sup>1</sup>, C. Marrone<sup>2</sup>, G. Mirizzi<sup>2</sup>, S. Garibaldi<sup>2</sup>, A. Rossi<sup>2</sup>, L. Ait-Alli<sup>3</sup>, P. Festa<sup>2</sup>, N. Assanta<sup>2</sup>, G. Santoro<sup>2</sup>

<sup>1</sup> Cardiology Postgraduate School, University of Pisa (Pisa)

<sup>2</sup> «Fondazione Toscana Gabriele Monasterio» (Massa, Pisa)

<sup>3</sup> National Council of Research - Institute of Clinical Physiology (Pisa)

### BACKGROUND

An integrated strategy that leads to early diagnosis of Tetralogy of Fallot (TOF) sequelae in adulthood should combine a detailed morphological and functional assessment through advanced imaging and an electrophysiological evaluation of the potential arrhythmogenic substrates. This combined diagnostic approach is fundamental to identify the best candidates to invasive procedures, such as pulmonary valve replacement (PVR) and arrhythmias radiofrequency ablation (RFA).

### AIM

Herein, we present a series of 3 adult patients with repaired TOF (rTOF) who underwent a percutaneous combined electrophysiological and hemodynamic procedure.

### METHODS

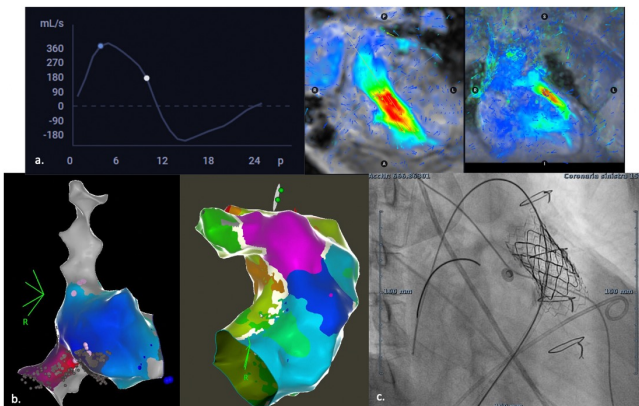
All patients underwent the following diagnostic algorithm: clinical and surgical history collection, EKG, Holter monitoring, NT-proBNP plasmatic level measurement, transthoracic echocardiography, cardiopulmonary exercise test and cardiac magnetic resonance (CMR). The risk score model from Ghoniem et al.<sup>(1)</sup> was used to stratify the annual risk of mortality. Two patients with high and one with moderate-to-high risk were candidate for a combined invasive procedure of electrophysiological study (EPS) with RFA followed by PVR.

(1) JACC Cardiovasc Imaging. 2022 Feb;15(2):257-268.

**PATIENT 1** was a 64-year-old woman with rTOF, admitted to hospital for right heart failure. CMR showed dilated right ventricle (RV) and a regurgitant fraction (RF) of 48% at the level of pulmonary valve (PV). The EPS documented a cavo-tricuspid isthmus (CTI)-dependent intra-atrial re-entrant tachycardia (IART), which was ablated. The electro-anatomical mapping (EAM) of the RV showed two of the classical anatomical isthmuses (AIs), specifically 1 and 4 (RV outflow tract patch to tricuspid annulus and ventricular septal defect patch to tricuspid annulus, respectively) with preserved conduction velocity ( $> 0,5$  m/s); accordingly, programmed ventricular stimulation (PVS) was negative for inducible arrhythmias. During that same procedure, a transcatheter balloon-expandable PV was successfully implanted.

**PATIENT 2** was a 37-year-old woman with double outlet RV (Fallot type) and a long history of surgical procedures, the last being the implantation of a RV-to-pulmonary artery conduit (Hancock 24). She was symptomatic for palpitations, with frequent premature ventricular complexes at Holter monitoring. CMR demonstrated dilated right chambers, an initial biventricular systolic dysfunction and a steno-insufficiency of the Hancock conduit. The EPS revealed a pathological right ventricular outflow tract (RVOT) and two slow-conducting AIs, which were prophylactically targeted through radiofrequency, since PVS was negative even before the ablation. Following conduit angiography and a sizing balloon test, a transcatheter balloon-expandable PV was implanted without complications.

**PATIENT 3** was a man with rTOF, lost at follow-up after surgical repair. Since early adulthood, he started to feel palpitations and frequent premature beats. At the age of 47, he was admitted to the emergency room due to a wide complex tachycardia, treated with antiarrhythmic drugs. CMR showed a dilated RV, an aneurysmatic RVOT and a severe pulmonary regurgitation. The EPS induced a right atriotomy-dependent IART, which was ablated, as well as the CTI. The EAM of the RV excluded the presence of slow conducting AIs and PVS was negative. During that same procedure, a RVOT occlusion test was performed, showing a complete occlusion with a 40 mm balloon without coronary compression. Because of a dilated outflow tract, the patient was scheduled for PVR with a self-expandable prosthesis, which was designed to adapt to relatively large RVOTs.



PATIENT 1: (a.) Phase-contrast quantification and 4D-flow MRI study of pulmonary regurgitation. (b.) Electro-anatomical mapping of right atrium and right ventricle. Radiofrequency ablation of cavo-tricuspid isthmus. (c.) Fluoroscopy image after the implantation of a transcatheter Melody® pulmonary valve.

### CONCLUSIONS

Our initial experience with combined invasive electrophysiological and hemodynamic treatment of patients with rTOF demonstrates that this interventional procedure is feasible and free from short-term complications.

A multiparametric assessment is mandatory before the procedure and high-risk patients may benefit from it.