CASE PRESENTATION

Cardiac resynchronization therapy in a child with tetralogy of Fallot

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Abstract: Cardiac resynchronization therapy is a common option in adult patients with heart failure and conduction abnormalities. Specific selection criteria for pediatric population are lacking. We report the case of a 14-year-old boy with significant pulmonary regurgitation subsequent to corrected tetralogy of Fallot and single chamber permanent pacing which presented with heart failure symptoms. Careful management was planned by a Heart Team including pediatric cardiologist, cardiac surgeon, interventional cardiologist and anesthesiologist. After undergoing cardiac surgery with complete correction of the pulmonary regurgitation and upgrade to a triple chamber pacing, the patient was discharged in good clinical status.
Keywords: cardiac resynchronization therapy, heart failure, pediatrics.

Rezumat: Terapia de resincronizare cardiacă este o opțiune frecventă la pacienții adulți cu insuficiență cardiacă și tulburări de conducere. Criteriile specifice de selecție pentru populația pediatrică lipsesc. Raportăm cazul unui băiat în vârstă de 14 ani cu insuficiență pulmonară semnificativă secundară unei tetralogii Fallot corectate și cardiostimulare permanentă monocamerală, care prezenta simptome de insuficiență cardiacă. Managementul pacientului a fost planificat cu grijă de o echipă formată dintr-un cardiolog pediatru, chirurg cardiac, cardiolog intervenționist și anestezist. După ce a fost supus unei intervenții chirurgicale cardiace cu corectarea completă a regurgitării pulmonare și trecerea la stimulare tricamerală, pacientul a fost externat în stare clinică bună.
Cuvinte cheie: terapie de resincronizare cardiacă, insuficiență cardiacă, pediatrie.

INTRODUCTION

Cardiac resynchronization therapy (CRT) has proven its effectiveness in appropriately selected candidates and became a standard of care in adult heart failure (HF) patients with conduction abnormalities. Current guidelines have specific recommendations regarding the selection of the CRT candidates, considering the clinical status, medical therapy, ECG and left ventricular ejection fraction (LVEF)[1].

Whether pediatric population might benefit from this invasive therapy or which are the selection criteria is not clear. All the big CRT studies have enrolled only adult patients[2]. In children, data come only from case reports or small case series. It is probably due to the lower incidence of heart failure in this age group, commonly limited to those with congenital heart disease[3-6].

CASE REPORT

We report the case of a 14-year-old boy with corrected tetralogy of Fallot and permanent cardiac pacing (single chamber pacemaker, VVIR) due to postoperative complete atrioventricular (AV) block which was referred for exertional dyspnea and growth retardation. The patient underwent cardiac surgery with complete correction (ventricular septal defect correction with

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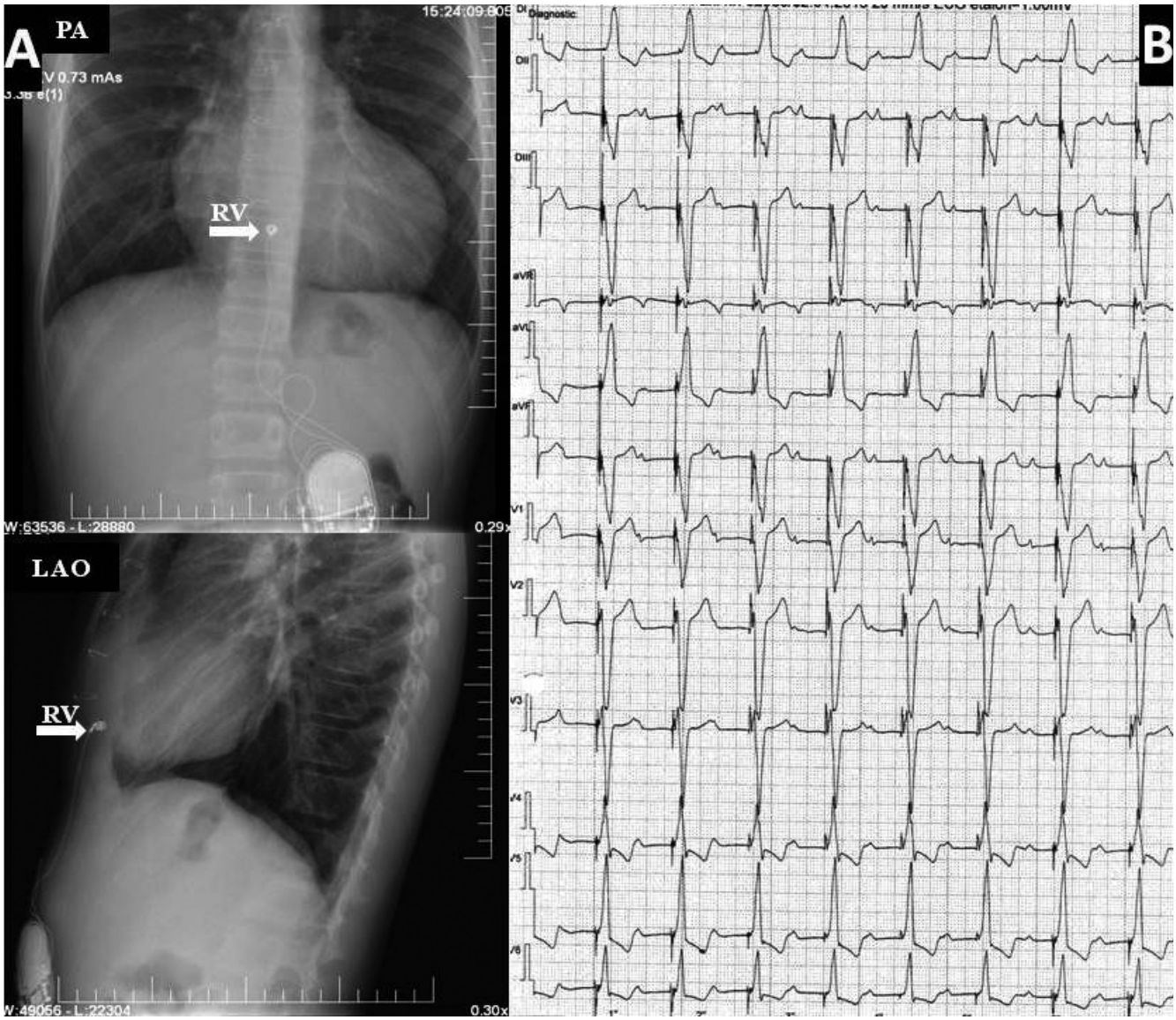

Figure 1. Single chamber cardiac pacing with epicardial lead on the right ventricle in tetralogy of Fallot with severe pulmonary regurgitation. Left panel (A): Chest X-ray. Right panel (B): Electrocardiogram. RV= right ventricular lead; PA=postero-anterior view; LAO=left anterior oblique.

pericardial patch, infundibular resection and transannular patch) during early childhood (7-months-old). Due to subsequent complete AV block he received a single chamber rate adaptive pacemaker with epicardial lead placed on the right ventricular (RV) free wall (Figure 1A). Six years later the pacemaker was replaced with a similar device due to battery depletion. At that moment echocardiography revealed significant pulmonary regurgitation without right heart dilatation, and paradoxical movement of the septum due to RV pacing. After five years, the pacemaker reached ERI (elective replacement indicator) and a new device was implanted. Echocardiographic evaluation showed dilatation of the right chambers of the heart. Surgical correction of the pulmonary regurgitation was proposed but the patient delayed therapy. Three years later, he was referred to our center for the reasons mentioned above. ECG showed sinus rhythm with AV dissociation and paced QRS complexes (QRS width=200ms with left bundle branch block (LBBB) morphology, Figure 1B). Compared to the previous echo examination, the right chambers were more dilated, the mechanical dyssynchrony worse (Figure 2A) and the left ventricular (LV) stroke volume reduced. Considering the clinical status and the progressive dilatation of the right heart, the patient underwent

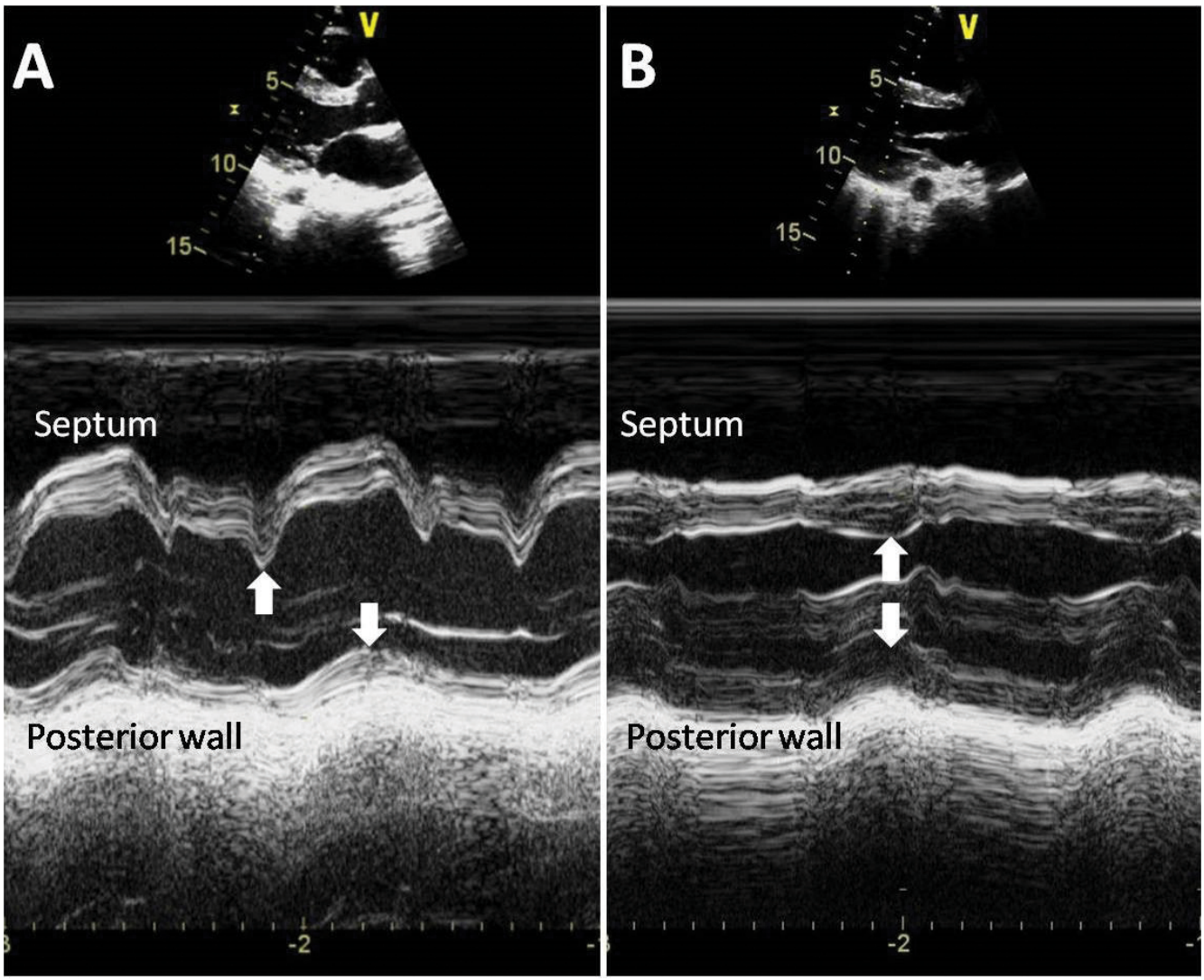

Figure 2. M-mode echocardiography in parasternal long axis view (PLAX) showing dyssynchronous/synchronous contraction of the interventricular septum and posterior wall (white arrows). Left panel (A): before therapy. Right panel (B): after therapy.

cardiac surgery with implantation of a biological prosthesis (Carpentier-Edwards Perimount Magna Ease) in pulmonary position (Figure 3A). During the same procedure 2 additional epicardial leads were placed: one on the right atrium and the other on the LV. Those, alongside with the RV lead were connected to a triple chamber cardiac pacemaker (Figure 3A). The device was programmed DDD biventricular mode with LV anticipation (Fig 3B). The postoperative echocardiography illustrated synchronous contraction of both septum and LV postero-lateral wall (Figure 2B) with improved stroke volume. After a few days the patient was discharged in good clinical condition.

DISCUSSION

The patient presented is particular due to its complex cardiac condition that led to HF at a young age. Significant pulmonary regurgitation subsequent to complete correction of the tetralogy of Fallot (with transannular patch) and the deleterious RV pacing were responsible for the development of the pathological condition. During surgery both conditions were corrected leading to improved cardiac function. Decision to perform upgrade from RV pacing to CRT in a pacemaker dependent pediatric patient was based on the ECG findings (QRS=200ms with LBBB morphology) associated to severe septal „bounce" on echocardio-

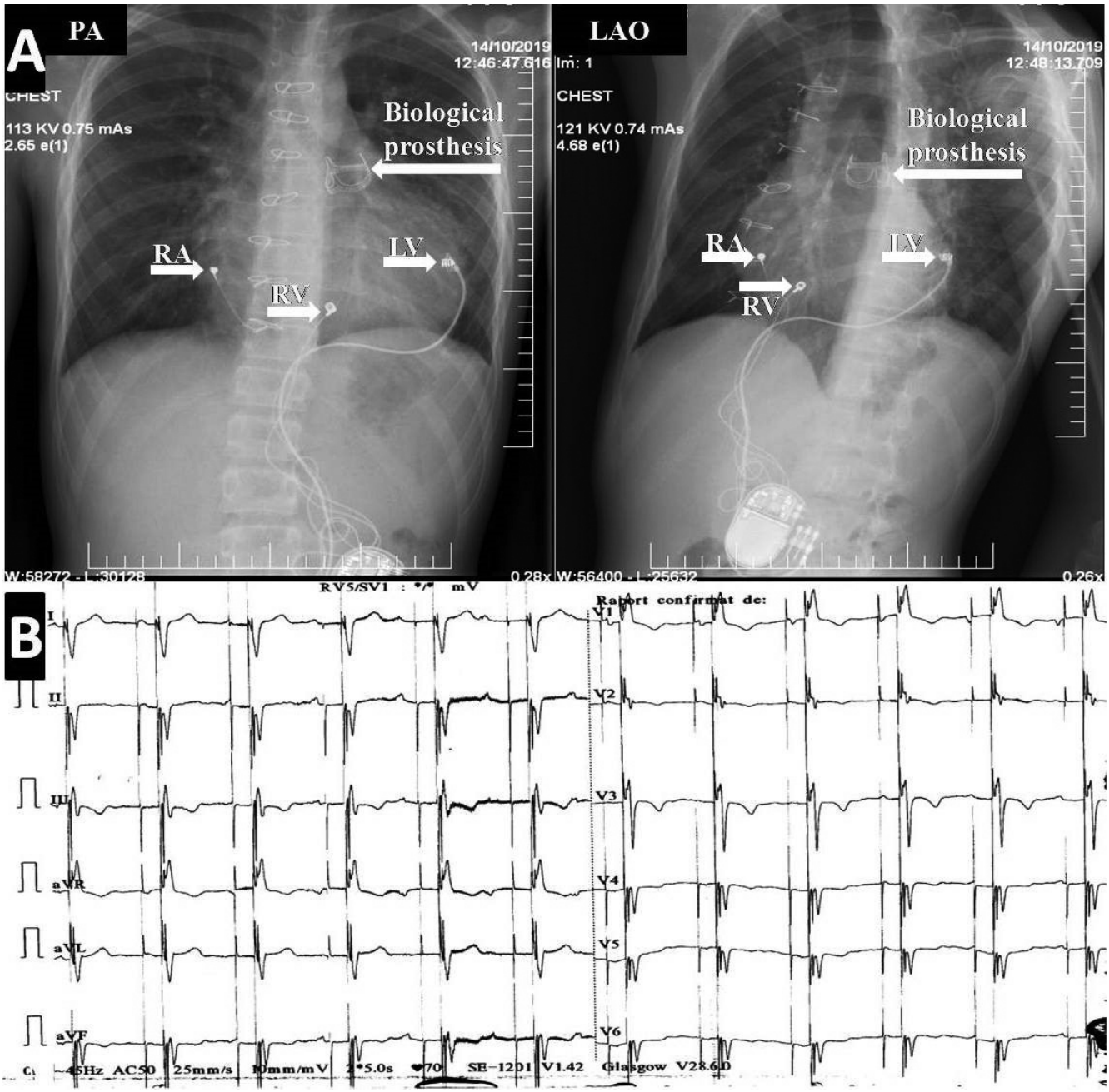

Figure 3. Cardiac resynchronization therapy with epicardial leads and Contegra graft in pulmonary position in repaired tetralogy of Fallot. Upper panel (A): Chest X-ray. Lower panel (B): Electrocardiogram. RA= right atrial lead; RV= right ventricular lead; LV= left ventricular lead; PA=postero-anterior view; LAO=left anterior oblique.

graphy. The echo exam was repeated after surgery while pacing either RV or both ventricles. The septal "bounce" was found only during RV pacing. This finding suggests that responsible for the mechanical dyssynchrony was mainly the non-physiological cardiac pacing. However, we cannot underestimate the role of pulmonary regurgitation in the development of HF, especially in the context of dilated right heart cavities.

Cardiac pacing in pediatric population is usually confined to complete congenital or acquired AV block. The later develops after the surgical repair of a congenital heart disease. Tetralogy of Fallot is most commonly involved[7]. Its correction is a two-step procedure consisting of closure of the ventricular septal defect with a pericardial patch, infundibular resection and, usually, transanular patch. The later results in pulmonary regurgitation with increased RV preload, leading

eventually to dilatation of the right heart chambers and HF. Once the RV dilates, it is recommended to treat pulmonary regurgitation either surgically (with a biological prosthesis) or interventional (transcatheter valve implantation) in order to avoid RV failure[8].

Pediatric patients with complete AV block do usually undergo single chamber pacemaker implantation with epicardial lead attached to the RV wall[9]. Kim et al have shown that up to 10% of patients developed HF after 10 years of RV pacing due to the atrioventricular and interventricular dyssynchrony[10]. There is a growing body of literature suggesting that development of pacemaker induced cardiomyopathy is highly dependent on the lead position. LV apical/midwall pacing appears to preserve better the cardiac function compared to RV pacing[9].

CRT is increasingly used for adult patients with HF and reduced ejection fraction who have conduction abnormalities. Restoration of both atrioventricular and interventricular synchrony usually results in improved cardiac performance and functional capacity[1,2]. However, up to one third of patients who undergo CRT according to current guideline criteria are non-responders to the therapy. New parameters for patient selection and CRT optimization are still under research[11,12].

Specific criteria for pediatric population are lacking. There are not too many children meeting the CRT criteria from adults. First of all, NYHA functional class was designed for adults. It is not clear if it's suitable for children considering the particular metabolism and physical activity that varies even between age groups. Secondly, QRS duration is changing with age. As a consequence, it is not unusual to have mechanical dyssynchrony even when QRS <120ms. Thirdly, adults do usually have LBBB QRS morphology, while in pediatric population right bundle branch block morphology is prevalent. This is due to the fact that children with HF have frequently a congenital heart disease. Moreover, in this specific subgroup, evaluation of the LVEF is more difficult due to the complex anatomy[2-6].

Conflict of interest: none declared.

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