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Case report

Cardiac arrest in patient with significant pulmonary regurgitation after surgical valvulotomy at 10 years of age for isolated pulmonary stenosis

Petr Klofáč^{*}, Tomáš Roubíček, Rostislav Polášek

Kardiocentrum, Krajská Nemocnice Liberec, a.s., Czech Republic

ARTICLE INFO

Article history:

Received 24 July 2017

Received in revised form

26 October 2017

Accepted 2 November 2017

Available online xxx

Keywords:

Congenital heart defect

Pulmonary stenosis

Pulmonary regurgitation

Reoperation

ABSTRACT

The number of adult patients with a congenital heart defect has increased. They are 2–3 times more numerous than children suffering from congenital heart defects, therefore, it is important to be aware of the most frequent congenital heart defects in adulthood – atrial septal defect, ventricular septal defect, aortic coarctation, tetralogy of Fallot and pulmonary stenosis. These patients either underwent one or more operations in childhood, or were not operated at all (defect was not significant or inoperable), or the heart defect was not diagnosed (mostly atrial septal defect). Some of the patients for various periods of time stop attending regular follow-ups and being asymptomatic (even in cases of hemodynamically significant defects), they do not seek medical attention. We present the case of a 46-year-old man with gradually progressing pulmonary regurgitation after surgical valvulotomy at 10 years of age. Despite regular follow-ups by the cardiologist, the patient was never referred to a specialized centre and cardiac arrest caused by ventricular fibrillation occurred after physical exercise.

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Introduction

Pulmonary stenosis represents 6% of all congenital heart defects in childhood. If isolated pulmonary stenosis is treated by surgical valvulotomy or by balloon valvuloplasty, there is a risk of significant pulmonary regurgitation in adulthood [1].

Case report

A 46-year-old patient underwent surgical valvulotomy for pulmonary stenosis at the age of 10 years old in Motol Faculty

hospital. In adulthood, the asymptomatic patient was followed by an outpatient cardiologist and had repeatedly echocardiographic examinations. Despite the fact that the echocardiography examinations showed a moderate pulmonary regurgitation and dysfunctional right ventricle (RV), the patient was not referred to a specialized centre for congenital heart defects. Later on, the patient was transported to our cardiology centre after cardiopulmonary resuscitation for OHCA (Out of Hospital Cardiac Arrest) caused by ventricular fibrillation during downhill skiing. The patient was defibrillated in total five times and spontaneous circulation was restored within 25 min. The history of the congenital heart defect was not known. The initial electrocardiogram (ECG) during admission showed atrial fibrillation with a right bundle

^{*} Corresponding author at: Kardiocentrum, KN Liberec a.s., Husova 10, Liberec 460 63, Czech Republic.

E-mail address: petr.klofac@nemlib.cz (P. Klobáč).

<https://doi.org/10.1016/j.crvasa.2017.11.002>

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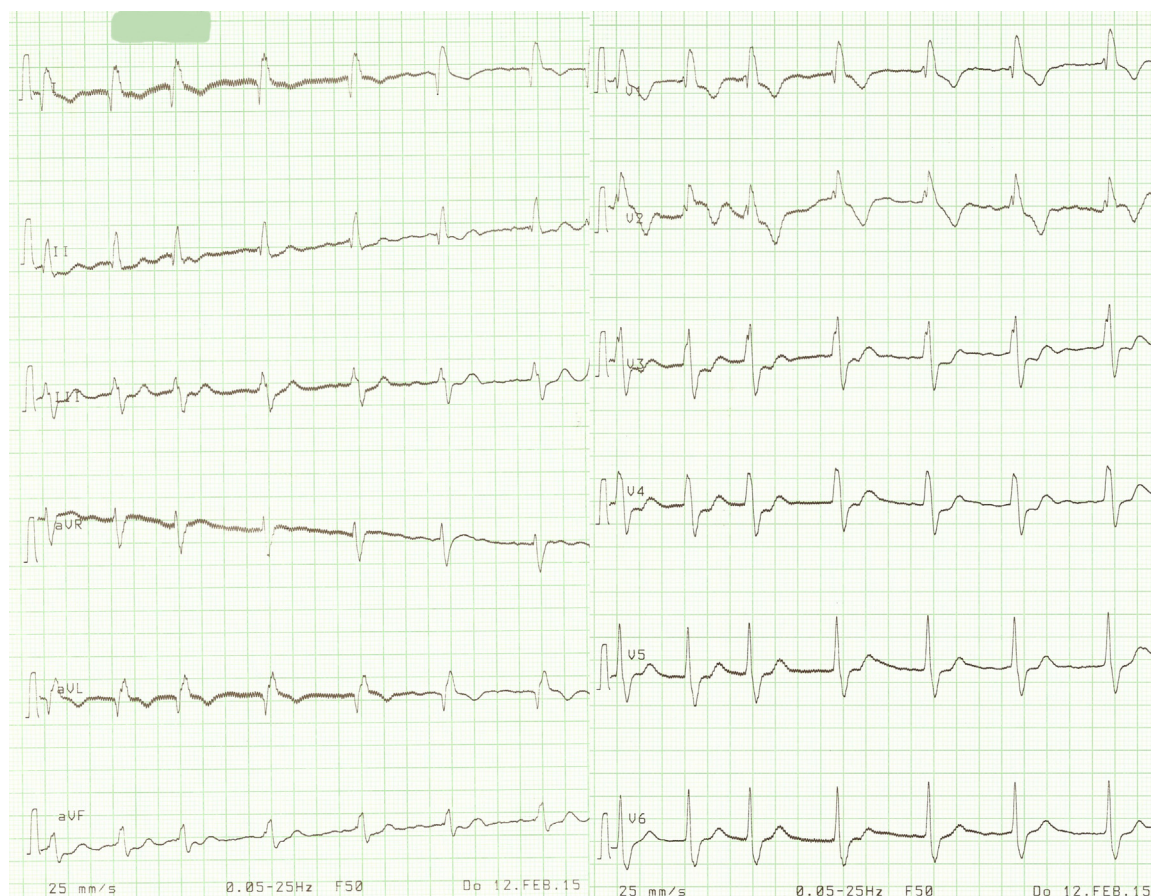


Fig. 1 – The initial ECG – atrial fibrillation and right bundle branch block.

branch block (RBBB) with 140 ms wide QRS (Fig. 1). After the admission to the coronary care unit, an echocardiography examination revealed a dilated and dysfunctional RV (Fig. 2), significant tricuspid regurgitation 4/4 and severe pulmonary regurgitation 4/4. These findings led to the suspicion of pulmonary embolism and as a result systemic thrombolysis was administered. Subsequently, CT angiography was performed and pulmonary embolism was excluded. The patient eventually disclosed the history of pulmonary stenosis and transesophageal echocardiography (TEE) confirmed significant pulmonary regurgitation (Figs. 3–5) as the reason of the dilatation and dysfunction of the RV and thus cause of cardiac arrest. NT-proBNP was 402 pg/l. The patient was then transported to the Department of Cardiac Surgery at Na Homolce Hospital. Cardiac surgeons for adult and paediatric patients together performed a pulmonary valve replacement (PVR) using a bioprosthesis and at the same time tricuspidal valve repair. Extracorporeal circulation lasted 100 min. Then they performed programmed ventricular stimulation and after the induction of sustained monomorphic ventricular tachycardia (only one morphology was inducted) a cardioverter-defibrillator (ICD) was implanted. Mapping and ablation of the ventricular tachycardia substrate in the right ventricle has not been performed yet, due to the likely complexity of the ablation in the field of remodelled RV. Two years after surgery, the patient is in good condition, has greater performance than

before surgery and starts to exercise again (although he was without symptoms before). Echocardiography shows good function of the pulmonary bioprosthesis (without noticeable regurgitation) (Fig. 6), only a mild tricuspid regurgitation 1/4 along the ICD electrode and persisting moderate dysfunction of the mildly dilated RV (Fig. 7). S' wave velocity of the tricuspid annulus is 9 cm/s and E/E' is 11. End-diastolic area of the right ventricle (RV EDA) has reduced from 50 cm² to 33 cm² and end-systolic area of the right ventricle (RV ESA) has reduced from 37 cm² to 23 cm². Fractional area change (FAC) has improved from 0.25 to 0.30. On the ECG the RBBB persists (Fig. 8), but with a more narrow QRS complex – 120 ms (against 140 ms before surgery) suggesting an improvement of the RV function. So far, the ICD has not detected any episodes of ventricular tachycardia or fibrillation.

Discussion

A congenital heart defect is a morphological anomaly of the heart and/or great vessels that is present since birth. The estimated incidence of congenital heart defects is reported as 6–12 for every 1000 live births. The defect may manifest itself at any time during life. Thanks to the excellent results of paediatric cardiac surgery and intensive care, 85–95% of patients live to adult age. Many patients with congenital

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