Heart transplantation due to advanced chronic heart failure after complete correction of tetralogy of Fallot

Transplantacija srca zaradi napredovalega srčnega popuščanja po kompletni korekciji tetralogije Fallot

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Izvleček

Predstavljamo 52-letnega bolnika več deset let po kompletni korekciji tetralogije Fallot. Glavni kasni zaplet je bilo srčno popuščanje zaradi odpovedi levega prekata. Transplantacija srca je bila edina terapevtska možnost.

Abstract

A 52-year-old patient decades after complete correction of tetralogy of Fallot is presented. The main late complication was left ventricular failure with consequential congestive heart failure. Heart transplantation was the only therapeutic option.

Introduction

Late complications seen in adult patients after complete correction of tetralogy of Fallot are: residual pulmonary regurgitation with consequent dilatation and failure of the right ventricle, residual right ventricular outflow tract obstruction, residual ventricular septal defect, aortic regurgitation with or without aortic root dilatation, supraventricular and ventricular arrhythmias and sudden cardiac death. Left ventricular dysfunction can be seen only occasionally and its aetiology is still not well understood. A variety of contributing factors, such as inadequate myocardial protection during previous repairs, chronic left ventricular overload due to longstanding palliative arterial shunts and/or residual ventricular septal defect, injury to anomalous coronary artery or longstanding cyanosis before repair, are suspected of triggering left ventricular failure.[1–3]

The majority of late complications can be surgically or percutaneously resolved. The management of heart failure in adult patients with congenital heart disease is still challenging and there are no guidelines for this particular group of patients.[4]

A case of advanced heart failure treatment in an adult patient after complete correction of tetralogy of Fallot in his childhood is presented.

Case report

A 52-year-old male was admitted to our hospital for progressive congestive heart failure late after complete repair of tetralogy of Fallot. At eight years of age, the patient underwent complete correction of the anomaly. He had been asymptomatic until the age of 39. At that time he developed an atrioventricular block. He was given a pacemaker. In the next two years he experienced several episodes of sustained ventricular ta-

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chycardia. Radiofrequency catheter ablation was successful only for a short time and an implantable cardioverter defibrillator was implanted. The patient was checked at yearly follow-up visits. Significant pulmonary regurgitation or obstruction of the right ventricular outflow tract has never been noticed. A nonsignificant residual ventricular septal defect was detected. The aortic regurgitation due to aortic dilatation was just mild. In the beginning we noticed mild left ventricular dysfunction, but no right ventricular dysfunction. In the next years, the patient gradually developed symptoms, including exertional dyspnoea, orthopnoea and fatigue. The patient was put on a regimen of an angiotensin-converting enzyme inhibitor, a beta blocker and low-dose furosemide. At the annual follow-up visits, the patient's condition was quite stable; he improved to New York Heart Association class II until his 51st year. Later on the symptoms of heart failure became more pronounced and he developed leg oedema and ascites. His hospitalizations for congestive heart failure were more and more frequent. Medical therapy with levosimendan was effective only for a short time. The patient was put on the emergency list for heart transplantation. His heart transplantation was successful and he underwent uneventful postoperative recovery.

The *electrocardiogram* on his last admission before heart transplantation demonstrated a regular pacemaker rhythm of 60 beats per minute with frequent ventricular ectopic beats. *Echocardiography on admission* revealed an enlarged left ventricle (EDD 66 mm, ESD 53 mm) with global hypokinesia and an ejection fraction of 30 % assessed by the Simpson method, and tissue Doppler velocities on mitral annulus were low (S_m 5.1 cm/s, E_m 6.2 cm/s) . The right ventricle was severely enlarged (basal 59 mm, mid 49 mm, base-to-apex 108 mm) with mildly to moderately depressed systolic function and low tissue Doppler velocities on the tricuspid annulus (S_t 10.8 cm/s, E_t 11.5 cm/s), and TAPSE was 17 mm. The transmitral Doppler patterns were consistent with a restrictive pattern of the left ventricle (transmitral E-wave and A-wave velocities of 0.72 m/s, and 0.31 m/s, respectively). The mitral valve was normal with mild functional regurgitation. Functional tricuspid regurgitation was severe with a retrograde velocity of 2.98 m/s. Both atria were enlarged (right atrium 67 mm, 70 mm, left atrium 63 mm, 76 mm). The pulmonary regurgitation was mild, without residual obstruction of the right ventricular outflow tract or the valve. The residual ventricular septal defect had been noted ever since the complete correction of the anomaly and assessed as a mild left-to-right shunt with high Doppler velocities through the shunt (up to 5.2 m/s). The ascending aorta was dilated at the sinus level to 44 mm. The aortic valve was morphologically normal and mildly insufficient. The time velocity integral of the left ventricular outflow tract was 11.6 cm.

Heart catheterization revealed moderate post-capillary pulmonary hypertension (pulmonary artery pressure 46/26 mm Hg, pulmonary capillary wedge pressure 22 mm Hg), elevated right ventricular end-diastolic pressures (22 mm Hg) and right atrial mean pressure (20 mm Hg). The coronary angiogram was normal.

Pathological findings. The explanted heart heart was enlarged, weighing 590 gr, with hypertrophy of the left ventricle and disseminated foci of fibrosis in the muscle wall. Histologically, there was myocyte hypertrophy, mild to moderate interstitial fibrosis and disseminated foci of replacement scars (Fig. 1).

Discussion

We presented an unusual course late after complete correction of tetralogy of Fallot. The surgical repair was excellent, without any significant typical late complications, except ventricular arrhythmias. The main late complication was left ventricular failure with consequential congestive heart failure and heart transplantation was the only therapeutic option. The development of symptoms of heart failure was gradual during fourteen years of follow-up. We excluded coronary and hypertensive heart disease as a causative reason for left ventricular failure. Left ventricular dysfunction in patients late after complete correction of tetralogy of Fal-

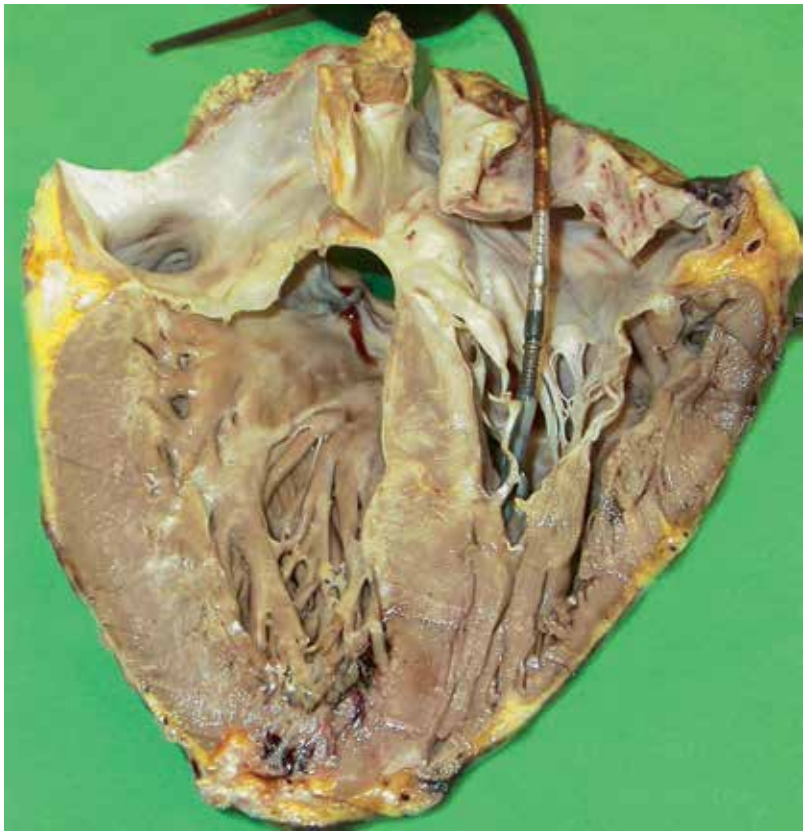

Figure 1: Macroscopically, the heart was enlarged, weighing 590 g, with hypertrophy of the left ventricle and disseminated foci of fibrosis in the muscle wall and interventricular septum. The implantable cardioveter defibrillator's lead is in the right ventricle.

lot is present only occasionally. It can be seen from a variety of factors. We can speculate that longstanding cyanosis before repair (eight years) and inadequate myocardial protection during surgical repair in the sixties in our patient could be the cause of heart failure later on. Some left ventricular overload due to residual ventricular septal defect and mild aortic regurgitation could be an additional factor to left ventricular failure. The sequence of events indicates that right ventricular failure was a consequence of left ventricular failure. Several pathophysiological mechanisms have been proposed to explain left ventricular failure after complete correction of tetralogy of Fallot. Autopsy and magnetic resonance studies have suggested that late ventricular dysfunction can be caused by myocardial fibrosis. The macroscopic and histological examination of the explanted heart confirmed hypertrophy of the left ventricle and disseminated foci of fibrosis in the muscle wall in our case. These findings are consistent with magnetic resonance studies in patients late after complete correction of tetralogy of Fallot.[5] The amount of fibrosis in adults with repaired tetralogy of Fallot is related to increased age, impaired exercise capacity, ventricular dysfunction and arrhythmia.[5,6] We can not explain the left ventricular dysfunction in our patient with left and right ventricular interaction because the right ventricular function was normal at the beginning.[7] Other possible mechanisms of decreased left ventricular function are also abnormalities of septal motion due to patching of ventricular septal defect or to septal fibrosis or myocardial injury in association with initial repair.[8] Longstanding cardiac pacing may also be associated with heart failure.[9]

In conclusion, isolated left ventricular dysfunction is a rare late complication after tetralogy of Fallot repair. We believe that isolated left ventricular dysfunction can be an independent entity after repair of tetralogy of Fallot without any known late residua. In advanced chronic heart failure, heart transplantation is the only reasonable option. Reports from literature about heart transplantation because of late complication in patients with tetralogy of Fallot are scarce.[10]

Conflict of interest and funding sources

None declared.

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