

Surgical Treatment of Arrhythmogenic Right Ventricular Cardiomyopathy with Absent Pulmonary Valve

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To the Editor: We described a successful treatment of a case diagnosed with arrhythmogenic right ventricular cardiomyopathy (ARVC) with absent pulmonary valve (APV).

A 10-year-old male child was referred to our emergency room for acute right heart failure who had two times of syncope before. On admission, the electrocardiogram showed that T-wave inversions were visible in most leads, QRS duration was prolonged with 120 ms, and the terminal depolarizations might be interpreted as epsilon waves in leads V1–V5 [Figure 1a arrows]. Transthoracic echocardiography and computed tomography scan were performed; the right ventricle chamber seemed larger than the left ventricle chamber [Figure 1b], with APV, atrial septal defect, and moderate tricuspid regurgitation.

On the basis of the diagnosis of ARVC combined with APV and for the frequent episodes of symptomatic dyspnea, the patient underwent pulmonary valve reconstruction and atrial defect closure. Fatty infiltration of right ventricle free wall and absent of pulmonary valve were detected during the operation and only inferior-posterior wall contained a little myocardial fiber [Figure 1c–1e]. In the situation with intact pulmonary annulus, three leaflets were reconstructed independently using autologous pericardium treated with 0.6% glutaraldehyde solution for 10 min [Figure 1f]. For the frequent episodes of syncope, the patient underwent implantation of a biventricular defibrillator.

The patient had been followed for 12 months and had experienced symptomatic improvement with return to the New York Heart Association Class I/II symptoms without any sustained ventricular arrhythmias or ICD discharges since surgery.

ARVC is a genetic form of cardiomyopathy characterized by fibrofatty replacement of predominantly right ventricular (RV) myocardium and is one of the leading causes of sudden unexpected cardiac death in young, apparently healthy individuals.^[1] As disease progresses, RV dilation and RV dysfunction may occur. The patients with unexplained syncope, non-sustained ventricular tachycardia, familial history of sudden death are potential candidates for Implantable Cardioverter Defibrillator (ICD) implantation even in the absence of ventricular arrhythmias.^[2] APV is a rare congenital heart disease with severe pulmonary insufficiency, characterized with aneurysmal dilation in the pulmonary artery. Because RV volume overload from significant pulmonary insufficiency may

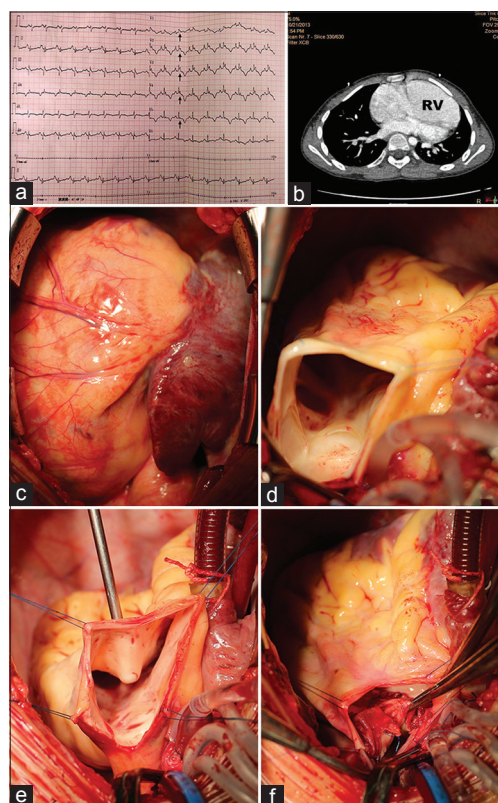


Figure 1: (a) Electrocardiogram showing that terminal depolarizations might be interpreted as epsilon waves in leads V1–V5 (arrows). (b) Computed tomography scan indicating the right ventricle chamber significantly enlarged. (c–e) The pathologic pictures showing fatty infiltration of right ventricle free wall and absent of pulmonary valve. (f) The leaflets were reconstructed independently using autologous pericardium in the situation with intact pulmonary annulus.

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in turn worsen RV function, surgical correction of pulmonary insufficiency could have therapeutic benefit. Symptoms usually appear between the ages of 30–50 years in ARVC patients and sudden death may be the first manifestation of the disease, mostly in previously asymptomatic young people and athletes.^[3] In case of older patients, the clinical presentation is mainly represented by signs and symptoms of right heart failure.^[4] Our case report showed in young patients, how this disease may present at the beginning with acute right heart failure, without ventricular arrhythmias but with episodes of symptomatic bradycardia that require the reconstruction of pulmonary valve and use of cardiac resynchronization therapy to avoid the well-known, long-term adverse effects of the right ventricular pacing.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Yan FP, Chen YY. A case of sudden death due to arrhythmogenic right ventricular cardiomyopathy. *Chin Med J* 2012;125:4318. doi: 10.3760/cma.j.issn.0366-6999.2012.23.031.
2. Ma KJ, Li N, Wang HT, Chu JM, Fang PH, Yao Y, *et al.* Clinical study of 39 Chinese patients with arrhythmogenic right ventricular dysplasia/cardiomyopathy. *Chin Med J* 2009;122:1133-8. doi: 10.3760/cma.j.issn.0366-6999.2009.10.003.
3. Romero J, Mejia-Lopez E, Manrique C, Lucariello R. Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC/D): A Systematic Literature Review. *Clin Med Insights Cardiol* 2013;7:97-114. doi: 10.4137/CMC.S10940.
4. More D, O'Brien K, Shaw J. Arrhythmogenic right ventricular dysplasia in the elderly. *Pacing Clin Electrophysiol* 2002;25:1266-9. doi: 10.1046/j.1460-9592.2002.01266.x.