Arrhythmogenic Right Ventricular Cardiomyopathy in Pregnancy

A Case Report and Review of the Literature

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Summary

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is predominantly a genetically determined heart muscle disorder that is characterized by fibro-fatty replacement of the right ventricular (RV) myocardium. The clinical spectrum of ARVC may represent from asymptomatic premature ventricular complexes to ventricular tachycardia (VT) and sudden cardiac death (SCD). It is a well-known leading cause of SCD in young adults.

There is no general consensus on the management of ARVC in pregnancy, and the preferred mode of delivery is uncertain. Herein, we report a case of ARVC diagnosed at 20 weeks of gestation following a sustained VT and treated with an implantable cardiac defibrillator (ICD). We also reviewed the current knowledge and approach to ARVC in pregnancy since the literature on this condition is based on case reports.

Key words: Implantable cardioverter defibrillator

ARVC is a primary disease of the myocardium characterized by fibro-adipocytic replacement of myocytes, mostly in the right ventricle. It should be suspected in a young or middle aged adult who has ventricular arrhythmias with left bundle-branch block (LBBB) morphology and does not have any other cardiac disease. Following the diagnosis of ARVC, the most important management decision is whether or not to implant an ICD for the treatment of sustained ventricular arrhythmias for prevention of SCD. It is now a standard practice for ARVC patients presenting with sustained VT and/or ventricular fibrillation (VF) to undergo placement of an ICD because of a high risk of recurrent VT and/or SCD.

The number of women with ARVC reportedly continues to increase and due to insufficient published data, it is rather difficult to evaluate the risk and management of ARVC during pregnancy and delivery. The diagnosis of ARVC based on development of VT during pregnancy and subsequent ICD implantation is a rarely seen situation.

Here we describe a patient diagnosed as ARVC at 20 weeks of gestation and managed with ICD placement during pregnancy.

Case Report

A 30-year-old woman (gravida 3, para 2), was admitted to Gynaecology and Obstetrics Hospital due to heart palpitations at 20 weeks of gestation. She had been asymptomatic until 20 weeks. When cardiac monitoring showed wide QRS complex regular tachycardia, she was transferred to our emergency department without any treatment by an ambulance in 10 minutes. She had no history of cardiovascular or any other disease. The patient was not on any medication but was taking supportive multivitamins. There was no history of smoking, but no alcohol consumption or illicit drug use. She had not experienced any problems during her previous pregnancies or deliveries. Her father had coronary artery disease but there was no family history of SCD among young adults.

In the admission period, she was hemodynamically stable. On physical examination, her blood pressure was 110/60 mmHg and heart rate was 200-210 bpm. Electrocardiography (ECG) showed sustained VT with inferior axis and negative concordance (Figure 1). In the emergency department, she was given adenosine 6 mg and then 12 mg intravenously (IV) in order to distinguish the tachycardia origin. However, sinus rhythm could not be restored so she was referred to us. The spontaneous sinus rhythm was established when we evaluated the patient. She was then admitted to the coronary intensive care unit.

Ventricular tachycardia (VT), at a rate of 200-210/min, that neither disturbed the hemodynamics nor caused any symptoms other than palpitations, occurred during admission to the coronary intensive care unit. Initially, 5 mg (IV) metoprolol was given followed by a second dose of 5 mg as sinus rhythm was not restored. Approximately 4 minutes after achieving a short period of sinus rhythm, VT at a rate of 190-200/min occurred again, causing no change in the hemodynamics. As VT was terminated and reoccurred again and again with the repeated dose of metoprolol 5 mg, amiodarone 150 mg (IV) was
Given. Instead of amiodarone, electrical cardioversion was considered, but a single dose of amiodarone was preferred for stabilizing the rhythm as the patient refused electrical cardioversion. Along with ongoing VT the patient reported feeling dizzy. Shortly afterwards she had a severe vomiting episode and then her blood pressure decreased notably. Subsequent to the vomiting attack, the VT was noticed to be terminated within seconds and the patient became hemodynamically stable. Once the sinus rhythm was re-established, the follow-up was uneventful.

A 12-lead ECG demonstrated normal sinus rhythm with frequent premature ventricular complexes (PVC) (3995/day), ventricular couplets (400/day), and triplets (30/day), and runs of non-sustained VT, up to 11 beats in duration (2 times/day, 11 salvos at maximum). Cardiac MRI presented the typical manifestation of ARVC indicating an enlargement of RV and right atrium without enlargement of the left ventricle and atrium; the RV was severely globally hypokinetic with lateral wall akinesia and the RV ejection fraction was 19% (Figure 3). The diagnosis of ARVC was based upon a set of major and minor criteria proposed by the International Task Force. She had two major criteria; 1) By MRI, her right ventricle was severely hypokinetic with lateral wall akinesia and the RV ejection fraction was 19%. 2) Her ECG demonstrated inverted T-waves in leads V1–V4 in the absence of complete RBBB. The patient also had one minor criterion; On Holter analysis there were 3995 ventricular extrasystoles per day. Thus, our patient fulfilled the latest criteria for definite ARVC diagnosis.

A fetal echocardiogram was performed at the 21st week of pregnancy and no abnormality was detected. Her other two children were also evaluated by echocardiogram and no abnormality was found. The relative benefits of ICD placement vs. antiarrhythmics for the prevention of SCD were discussed with the patient. After obtaining informed consent, a single chamber ICD was placed under local anaesthesia without any complications of note at 21 weeks of gestation. The abdomen of the patient was protected by a lead blanket. The patient tolerated the procedure well. The patient and fetus were monitored for 3 days after ICD implantation. In this period, numerous nonsustained VT episodes were observed and metoprolol was given to the patient to decrease her palpitations. A consultation with the Department of Obstetrics about the usage and dosage of metoprolol therapy revealed no contraindication to beta-blockers for her pregnancy. Finally, she was discharged on metoprolol (50 mg/day). She was monitored very closely for the remaining trimesters of her pregnancy. At 32 weeks of gestation, an echocardiogram demonstrated no changes in her cardiac functions and the device did not record any arrhythmias. She had no further problems during the rest of her pregnancy.

She delivered her baby in the 37th week of pregnancy by
caesarean section due to fetal distress (oligohydramnios) under spinal anaesthesia. The device was left off during delivery and no complications, such as arrhythmias, were observed during labour. The newborn was a healthy boy, with a birth weight of 3800 grams and APGAR score of 9/10 at the 1st and 5th minutes. Echocardiography of the baby was normal. After 3 days of hospitalization, she was discharged with metoprolol (50 mg/day) and continued breastfeeding for 6 months. The analysis of stored electrograms revealed no significant ventricular arrhythmia throughout the 6 months of ICD implantation.

**Discussion**

ARVC is characterized pathologically by myocardial atrophy, fibrofatty replacement, fibrosis and ultimate thinning of the wall with chamber dilatation and aneurysm formation. These changes consequently produce electrical instability precipitating VT and SCD. ARVC should be suspected in a young patient with palpitations, syncope, or aborted SCD. VT with LBBB morphology is the classic presentation. Other electrocardiographic abnormalities such as inverted T waves in right precordial leads (V1–V3) and frequent premature ventricular complexes (PVCs), even in asymptomatic patients, should arouse the suspicion for this cardiomyopathy. The main goal of therapy is to prevent serious events, which requires identifying high-risk patients for malignant arrhythmias and SCD.

The diagnosis of ARVC is based upon a set of major and minor criteria proposed by the International Task Force. Patients must either meet two major criteria, one major and two minor criteria, or 4 minor criteria to be diagnosed as ARVC. Two of the major and one of the minor diagnostic criteria for ARVC according to the new Task Force Criteria were present in our patient, ie, inverted T waves in leads V1–V4 in the absence of RBBB on ECG, severe hypokinesis of the right ventricle with lateral wall akinesia and reduced ejection fraction (19%) of the RV observed by MRI, and 3995 ventricular extrasystoles per 24 hours on ECG Holter examination.

Pregnancies with dilated and hypertrophic cardiomyopathies are common, but only a few cases of pregnancies with ARVC have been reported. Therefore, it is difficult to assess the risks of pregnancy and delivery in patients with ARVC. Furthermore, sustained VT developed during the pregnancy in only one of these cases. In the patient mentioned in this case, VT had developed following physical exercise at the 34th week of both her pregnancies. As sinus rhythm was achieved spontaneously approximately 20 seconds later, there was no need for medical or electrical cardioversion. In our case VT developed in the 20th week, during rest and continued for minutes. Sinus rhythm could be restored immediately by medical cardioversion.

During pregnancy, plasma volume, cardiac output and heart rate increase, hematocrit decreases, and physiologic anemia is established. VT may be triggered during pregnancy as a result of these hemodynamic changes. It is noteworthy that the published reports have found that most of these patients tolerate these conditional hemodynamic changes induced by pregnancy. Although pregnancy alone is a physiological stress factor, in our patient there was no other reason inducing the arrhythmia. Nevertheless, it has been reported that exercise itself can induce fatal arrhythmia in ARVC patients. The initiation of VT was associated with a typical catecholamine surge seen on exercise in the setting of the presence of arrhythmogenic substrate characteristic of these patients. Therefore, as exercise may increase heart rate in a pregnant woman with a diagnosis of ARVC, it is better to recommend to avoid exercise.

Considering the paucity of reported data in regard to the management of ARVC patients during pregnancy, it is still unknown whether an arrhythmic event could be preventable by any treatments. In a case reported by Iriyama, the propranolol treatment that the patient had been taking was not given during the pregnancy on her own request and VT occurred at the 34th week. Similarly in the case of Güdücü, the patient taking regular metoprolol and propafenone treatment for 3 years with the diagnosis of ARVC refused to use the drugs on her own request and she became symptomatic at the 32nd week. In her Holter ECG at the 32nd week, frequent PVC attacks were detected. In our case, the patient was discharged with metoprolol treatment after implantation of an ICD at the 21st week and she was asymptomatic until delivery. The analysis of stored electrograms revealed no significant ventricular arrhythmia at 6 months post ICD implantation under metoprolol treatment.

Treatment of ARVC is based on the prevention of arrhythmia with beta-blockers, especially with sotalol, which prevents arrhythmia caused by ARVC in 60% to 70% of patients. Propafenone was reported to be safe in the treatment of ventricular arrhythmia during pregnancy. Combination therapy (Class IC drugs plus beta-blocking drugs, or amiodarone plus beta-blocking drugs) frequently is effective in preventing recurrent VT. Unfortunately, amiodarone therapy during pregnancy (category D) may cause fetal/neonatal hypothyroidism and, less frequently, goiter. Thus, the use of amiodarone in pregnancy should be limited to VTs which are sustained, monomorphic, hemodynamically unstable, refractory to electrical cardioversion, or not responding to other drugs. In our case, although the hemodynamics were stable, by considering the fetal side effects, electrical cardioversion which is reliable in every phase of pregnancy might have been preferred instead of amiodarone. However, single dose-amiodarone was preferred because the patient refused the electrical cardioversion. Only a 150 mg-loading dose was given to the patient and a maintenance infusion dose could not be administered.

Prophylactic therapy with a cardioselective beta-blocking agent, such as metoprolol, may be effective. There is considerable experience with beta blockers and no teratogenicity has been reported to date, even if in some cases intrauterine growth delay and bradycardia have been reported. In our case, although oligohydramnios was detected at the 37th week of pregnancy, intrauterine growth retardation was not evident in the fetus and the baby was born healthy.

ICD use is recommended for patients who have a documented episode of sustained VT or cardiac arrest or who have high-risk features for SCD. Recently, ICD therapy for the prevention of SCD in patients with ARVC is becoming standard treatment. However, little is known regarding the outcome of pregnancy in women with ICD. We have found only one published case of a patient diagnosed with ARVC during her pregnancy and treated with ICD placement. In the last years, only a few studies of ICD therapy during pregnancy have found that...
the majority of women completed and tolerated pregnancy and delivered without serious complications. Thus, ICD placement should be considered in pregnant woman with ARVC to prevent episodes of SCD.

The preferred mode of delivery in women with ARVC is uncertain. Bauce, et al. reported that a caesarean section was performed in 4 of the 6 patients in whom the disease was considered to be more dangerous on the basis of both morphological changes and the history of ventricular arrhythmias. The remaining two cases with only isolated PVC or no arrhythmias underwent spontaneous vaginal delivery. Iriyama, et al. preferred to perform elective caesarean section due to a history of major arrhythmias. Our patient delivered by caesarean section due to fetal indication (oligohydramnios).

It remains unclear whether an ICD should be on or off during delivery. Recurrence of VT may result in decreasing placental perfusion due to maternal hypotension and could be dangerous for the fetus. In contrast, ICD shocks are a concern due to fetal indication (oligohydramnios).

We left the device turned off during caesarean section, since electrocautery was to be used. Our case is the first report of an ARVC patient who had a sustained VT that required medical cardioversion to be terminated during pregnancy. This is also the second case of a pregnant ARVC patient who underwent ICD implantation during her pregnancy. On the basis of other published studies and our case report, it can be said that the pregnancy is well tolerated in women affected by ARVC and can be managed successfully with close monitoring and antiarrhythmic drugs when necessary. ICD implantation also should be considered to prevent SCD during pregnancy to protect the life of the mother. Pregnancy seems not to cause an increase in major ICD-related complications. Despite all of these experiences, more case studies are needed to establish the most appropriate management of ARVC during pregnancy.

**REFERENCES**


