



VENTRICULAR TACHYCARDIA IN PATIENTS WITH ARRYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

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Abstract

Objective: This study was conducted to explore Ventricular tachycardia in patients with arrhythmogenic right ventricular cardiomyopathy

Materials and methods: A prospective study was conducted at Cardiac Electrophysiology Department, Hayatabad Medical Complex Peshawar Pakistan from 1st January 2015 till 31 December 2020, with 107 patients who suffered from sustained ventricular tachycardia (VT) being examined. Following etiological evaluation, 15 patients were found to have ventricular tachycardia (VT) causing ARVC as per the Modified Task Force Criteria. To better understand this entity in our study, we observed their clinical profile in detail. All the data was analyzed statistically.

Results: A total of 15 patients fulfilled the criterion for inclusion in our study. There were 12 (80%) male patients and 3 (20%) were female patients. Age of the patients ranged from 15 to 55 years whereas mean age at presentation was 30 years. 80% of the patients had abnormal ECG findings. 10 patients (66%) had repolarization abnormalities, and 5 (33%) had depolarization abnormality due to Epsilon wave interference. 9 patients who were hemodynamically stable were administered antiarrhythmic drugs. 7 patients reverted to the drugs only, and 2 needed further electric cardioversion due to sinus rhythm failure. Intravenous drugs restored the sinus rhythm with a 78% success rate. In six patients with hemodynamic instability, DC cardioversion was used to reverse the effects of a combination therapy that involved both medical and electric cardioversion. Antiarrhythmic drugs were administered to 9 patients, among them all 6 who successfully terminated VT. Amiodarone was used by 56% of the patients and sotalol was taken by 2 patients. Two patients were treated with a combination of different drugs, including amiodarone, sotalol, and lignocaine. An Automated implantable cardioverter defibrillator (AICD) was implanted in 9 patients.

Conclusion: Ventricular tachycardia frequently occurs in patients with arrhythmogenic right ventricular cardiomyopathy and may significantly contribute to morbidity and mortality

Keywords: Ventricular tachycardia; Arrhythmogenic right ventricular cardiomyopathy

Introduction

The hereditary variant of cardiomyopathy known as arrhythmogenic right ventricular cardiomyopathy (ARVC) is often passed down through an autosomal prevailing pattern. Fibrofatty replacement,

mainly of the right ventricle (RV) muscle, is its characteristic. While it mostly affects the right ventricle, it can also have an impact on the LV musculature. Biventricular heart failure (HF), potentially fatal ventricular arrhythmias, and sudden cardiac death (SCD) are possible outcomes. (1) of the young athletic population, it accounts for 11 to 22 percent of cases of sudden cardiac mortality. Giovanni Maria Lancisi, the physician to the Pope, initially discussed ARVC/D in his 1736 work *De Monty Cordis et Aneurysmatibus. Six*. Nonetheless, the disease's initial clinical profile was released in 1982.(2). Myocardial atrophy, fibrofatty replacement, fibrosis, and wall thinning with aneurysm development are its pathological characteristics. (3) Thirty to fifty percent of ARVC/D cases have been shown to have a genetic basis.(4) Variable penetration and incomplete expression are typical autosomal dominant traits that inherited diseases. The majority of the genes found encode the proteins in the desmosome family. ARVC/D has been linked to acquired factors as well. Arrhythmogenic cardiomyopathy and other viral myocarditis are the most strongly linked. ARVC/D patients' differing presentation may be attributed to genetic heterogeneity and modifying factors like exercise and viral myocarditis.(5.) Arrhythmias in the ventricles, particularly RV, are the most common clinical manifestation of ARVC/D. The arrhythmias can manifest as either isolated premature ventricular beats or sustained VT or vascular fibrillation, which may result in sudden cardiac death. Typically, the disease's symptoms are palpitations, chest discomfort, or being near syncope. The LBBB morphology of RV-derived arrhythmias is most commonly associated with sustained monomorphic (SUM) VT. In cases where the left ventricle is involved, right bundle branch block morphology (RBBB) can also occur. The diagnosis of ARVC/D is often challenging due to the lack of evidence to confirm or disprove the condition. To determine the diagnosis, one can rely on history, physical examination, and specific tests like ECG, EKG, CMR, or ventriculography. The Clinical Diagnostic Criteria for ARVC/D were published in 1994 by the Original International Task Force and evaluated the disease's structural, histological, ECG, arrhythmic, and familial characteristics. (6) the aim of the current study was to find out Ventricular tachycardia in patients with arrhythmogenic right ventricular cardiomyopathy

Materials and methods

A prospective study was conducted at Cardiac Electrophysiology Department, Hayatabad Medical Complex Peshawar Pakistan from 1st January 2015 till 31 December 2020.

Inclusion criteria Those who had sustained ventricular tachycardia (VT) and those with ARVC/D as proposed by the task force were included in the evaluation of any confirmed cases that had been admitted to the hospital or presenting to accident emergency department, and all others were assessed for their aetiology.

Exclusion criterion: Following patients were excluded

1) Those who experienced sustained VT had other causes, such as CAD, DCMP, and idiopathic associations. 2) Patients who died shortly after hospitalization, preventing them from receiving further evaluation. 3) Those who chose not to undergo further evaluation. Only the first episode was considered as an index event in patients with recurrent episodes of sustained ventricular tachycardia (VT).

Consent: Consent was granted to each subject upon detailed explanation of the study.

Initial Evaluation;

The "Brugada algorithm" and/or "The Avr Verecke Alogrithm" were used to diagnose sustained VT in every patient, who underwent a battery of tests after an acute abortive attack.(7,8). The hemodynamic status of patients presented was used to divide them into two groups: those that were stable and those who were unstable. Rapid cardioversion was necessary when loss of consciousness, hypotension, shock, or congestive heart failure necessitated hemodynamic instability. If not, the patients were deemed hemodynamically stable. Every patient was examined etiologically. The ECG

was observed in both the resting and VT phase. All patients underwent 2-D echocardiography. Cases were subjected to an exercise test and 24-hour Holter monitoring. Patients with suspected ARVC/D based on family history, ECG findings, echocardiography, stress test, or 24-hour Holter monitoring underwent a cardiac MRI analysis. Only those who met the Modified Task Force criteria of ARVC/D were considered for the ARV/ D group. (9)

Other relevant investigation.

Serum electrolytes

Serum potassium levels below 3.5mg/dl were deemed hypokalemia. Hypomagnesemia was defined as serum magnesium levels less than 1.5mg/dl.

VT termination and follow up

The treatment for ending VT was carried out in accordance with the current ACLS guidelines. Hemodynamically unstable patients were given DC cardioversion. Until the defibrillator was ready for DC shock, patients with pulseless VT were subjected to CPR. Patients who were hemodynamically stable were administered recommended doses of intravenous antiarrhythmic drugs. Visiting physicians were free to select their antiarrhythmic drug. In patients with unstable heart rhythms who did not respond to antiarrhythmic drugs, electrical cardioversion was given.

Results

A total of 107 patients having ventricular tachycardia were enrolled in this study and among them only 15 patients fulfilled the criterion of ARVC/D as proposed by modified task force criteria. There were 12 (80%) male patients and 3 (20%) were female patients. (Figure 1) Age of the patients ranged from 15 to 55 years whereas mean age at presentation was 30 years . Five patients with a positive family history of early sudden cardiac death (SCD) were identified. The percentage of Hemodynamic instability was 40 % (n=6) . The most common configuration was Monomorphic VT 87% (N=13) and 2 patients had Polymorphic VT. 11 patients (73%) had Left Bundle Branch Block (LBBB) morphology and 2 patients had Right Bundle Branch Block (RBBB) morphology. 80% of the patients had abnormal ECG findings. 10 patients (66%) had repolarization abnormalities, and 5 (33%) had depolarization abnormality due to Epsilon wave interference. All patients underwent echocardiography (Figure 3). A total of 11 patients (73%) had typical ARVC/D changes that met a major diagnostic criteria, while only 20% reported minor modifications. The results of the Cardiac MRI scan revealed changes that aligned with the major diagnostic criteria in 9 patients (60%). Our patient was not subjected to invasive tests such as right ventriculography and endomyocardial biopsy. 9 patients who were hemodynamically stable were administered antiarrhythmic drugs. 7 patients reverted to the drugs only, and 2 needed further electric cardioversion due to sinus rhythm failure. Intravenous drugs restored the sinus rhythm with a 78% success rate. In six patients with hemodynamic instability, DC cardioversion was used to reverse the effects of a combination therapy that involved both medical and electric cardioverted. Antiarrhythmic drugs were administered to 9 patients, among them all 6 who successfully terminated VT. Amiodarone was used by 56% of the patients and sotalol was taken by 2 patients (Figure 4). Two patients were treated with a combination of different drugs, including amiodarone, sotalol, and lignocaine. An Automated implantable cardioverter defibrillator (AICD) was implanted in 9 patients. (Figure 5). The presentation of two patients with hemodynamic instability resulted in mortality.

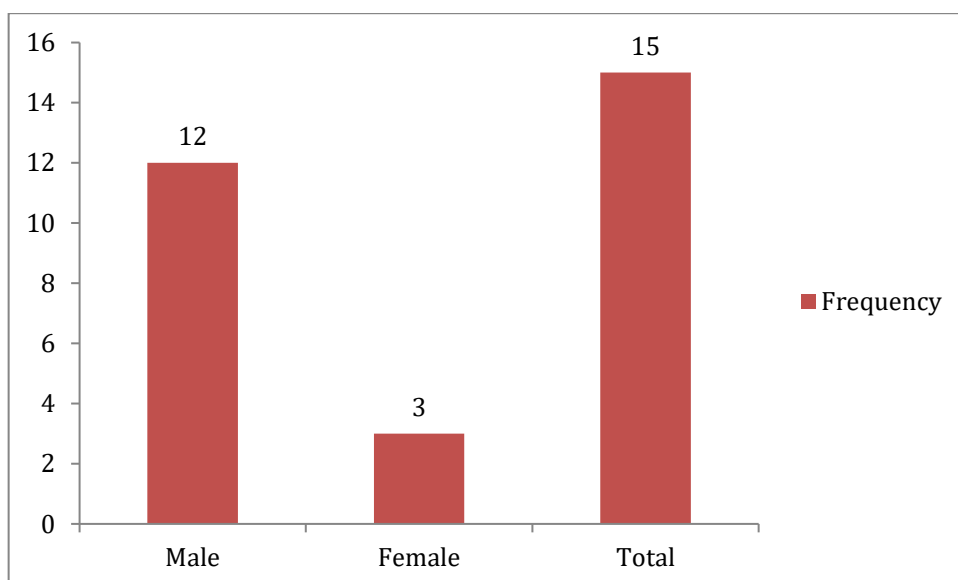


Figure 1: Gender wise distribution of patients

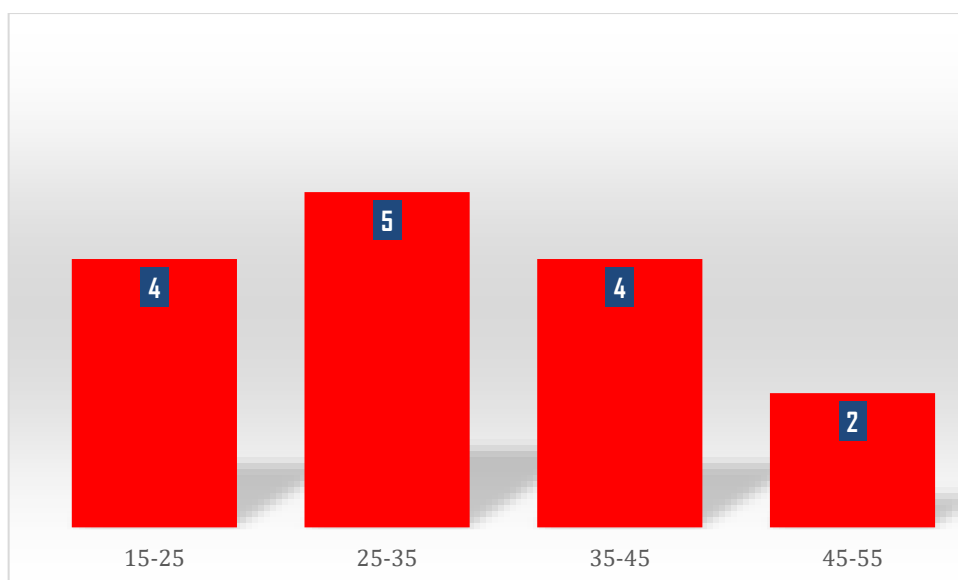


Figure 2: Distribution of patients based on age

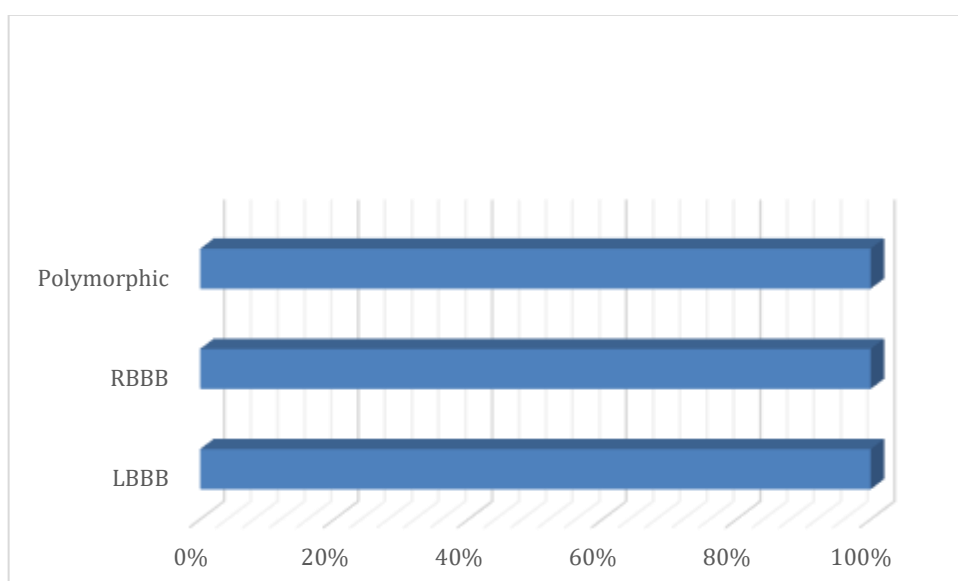


Figure 3: Showing patterns of ECG morphology seen in patients (%)

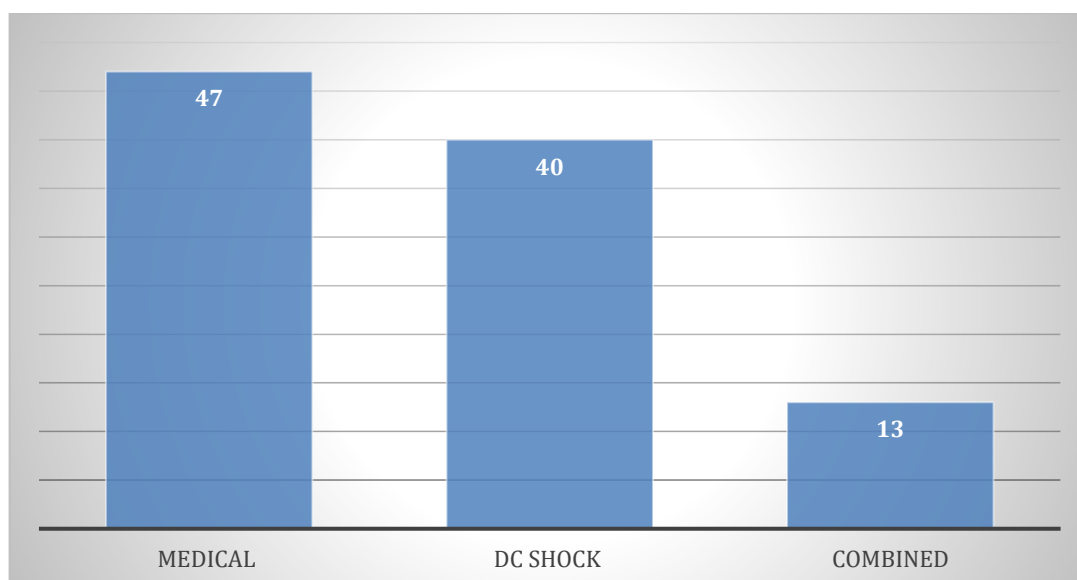


Figure 4: Showing treatment modality received by patients

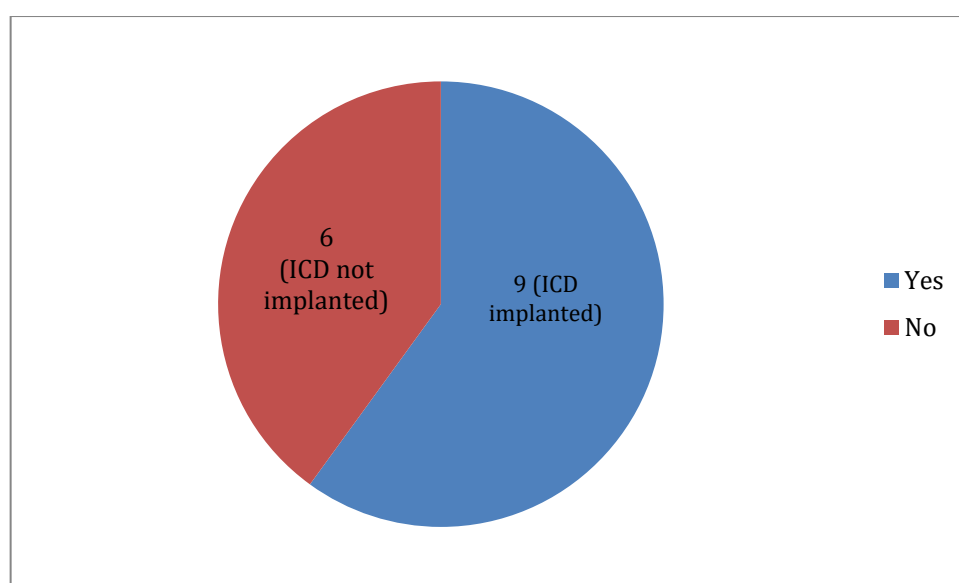


Figure 5: Frequency of patients with implanted ICD

Discussion

The patients who presented with ARVC/D as sustained VT were observed, leading to the following conclusions. Males are impacted more frequently than female. This condition occurs from an adolescent to an adult age. Most commonly the morphology on ECG is called LBBB. In many cases, hemodynamic instability is present and has a poor prognosis.

All of our patients received diagnosis without invasive testing, such as endo-myocardial biopsy and right ventriculography. Our study included 80% male patients, which is consistent with previous studies that found a higher incidence of ARVC/D in men.(10,11) Our patients aged 15 to 55 were present at the time of presentation. ARVC/D is most prevalent in 5 to 40 years according to the data.(12) The considerable difference in presentation age indicates that ARVC/D remains hidden for varying lengths of time in different individuals.(13) Your research investigated the presentation of ARVC/D patients as sustained VT, which is one of its most significant presenting symptoms.(14) The task force's modified criteria consider a family history of first-degree relatives of patients with onset-related sudden cardiac death (SCD) to be merely invasive diagnostic criteria. Our cohort's ECG findings indicated that the majority of sustained VT in 11 patients was due to LBBB morphology. This is in line with the data published on the same topic before.(15) The morphology of sustained VT was polymorphic and followed the LBBB path. The fact that both patients had haemodynamic

instability on presentation of polymorphic VT should be noted here. In their study, Domanovitis et al. found that the ECG characteristic of VT presented in the emergency department was only linked to the haemodynamic instability of patients with the condition. (16) ARVC/D can be easily and helpfully diagnosed using echocardiography. The imaging of structural and functional abnormalities in the right ventricle has been facilitated by this cost-effective, noninvasive technique. Measuring right ventricular function requires multiple locations, including the inflow and outward tracts, due to the disease's focus. (17,18). We conducted echocardiography on every patient in our study. Echocardiography suggests that ARVC/D is characterized by increased dilation of the right ventricle (RV) with localized aneurysms and dyskinesia in the inferobasal region. ARVC/D has been extensively studied using 2-D echocardiography, which is both highly specific and predictive. The majority (73%) of the patients had typical ARVC/D changes that met the major diagnostic criteria, while only 3 patients showed similar symptoms. All 15 patients underwent an MRI scan for their heart disease. According to quantitative analysis, ARVC/D patients exhibit significantly higher right ventricular end diastolic diameter and outflow tract area and RV ejection fraction than controls. (19,20). The cardiac MRI results in 9 patients were consistent with the major diagnostic criteria. Beta-blockers, lignocaine, sotalol, and amiodarone are among the drugs that have been studied to prevent lifelong arrhythmias of ARVC/D. (21) Our study utilized only amiodarone, sotalol, lignocaine, and I/V betablockers as antiarrhythmic drugs were not available. These drugs were administered either alone or in combination. Amiodarone was administered to 5 patients, and sotalol was used only by 2. Two more patients were treated with a combination of different drugs, such as amiodarone, sotalol, lignocaine, and i/v beta blockers. Patients who experience haemodynamic instability or unremittable conditions without reversible VT may still require the use of DC shock as a crucial treatment method. The ICD device can be used as a standard treatment to terminate life-threatening arrhythmias in patients with ARVC/D. ICD therapy is recommended for the prevention of sudden cardiac death (SCD) in patients with sustained VT or ventricular fibrillation (Class 1 recommendation), as well as for patients showing signs of extensive disease, a positive family history, or undiagnosed syncope (class 2a recommendation). (22) Our study examined patients with ARVC/D and sustained VT. Although ICD was recommended in all patients, it was only implemented in 9 patients for various reasons. The hospital experienced the death of two patients. The two patients were exhibiting hemodynamic instability when they presented.

Conclusion

VT frequently occurs in patients with ARVC /D and may significantly contribute to morbidity and mortality. Males are more frequently affected than females. Hemodynamic instability at the time of demonstration carries a poor prognosis

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