CLINICAL RESEARCH STUDY

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Incidence and Prognosis of Ventricular Arrhythmias in Patients with Congenital Left Ventricular Aneurysms or Diverticula

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ABSTRACT

BACKGROUND: Patients with congenital left ventricular aneurysms and diverticula may present with arrhythmia. The incidence of ventricular arrhythmias and the clinical outcome of these patients have not been reported to date.

METHODS: Among 250 consecutive patients with congenital left ventricular aneurysms and diverticula detected by echocardiography, the clinical outcome of patients who presented with ventricular arrhythmias or associated symptoms was investigated.

RESULTS: Of 250 patients with congenital left ventricular aneurysms and diverticula, 30 had ventricular arrhythmias or syncope at initial presentation. During a follow-up of 85 months, spontaneous ventricular tachycardia occurred in 17 of these patients (57%). Ventricular tachycardia was sustained in 13, with a monomorphic pattern in 9 patients. In 82% (11 patients), ventricular tachycardia was inducible during electrophysiologic testing. In 7 patients a sustained monomorphic ventricular tachycardia with a right bundle branch block pattern similar to the clinical tachycardia was induced. Twenty patients were treated with antiarrhythmic agents. Eleven patients received an implantable cardioverter defibrillator. Appropriate device discharges were observed in 73% during a follow-up of 61 months. One patient underwent surgical resection of a congenital left ventricular aneurysm. Three patients underwent successful catheter ablation for incessant ventricular tachycardia. Of these, 2 were free of any clinically relevant arrhythmia during follow-up. Three patients died (10, 41, and 89 months after initial presentation). In 2 of them, the cause of death was attributed to ventricular arrhythmia.

CONCLUSION: The clinical outcome of patients with congenital left ventricular aneurysms and diverticula and arrhythmia is variable. Clinical ventricular tachycardia in these patients is often monomorphic and usually inducible during electrophysiologic study, indicating a role for this test in risk stratification. Appropriate discharges are frequent in implantable cardioverter defibrillator recipients with congenital left ventricular aneurysms and diverticula.

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Congenital left ventricular aneurysms and diverticula are rare disorders that seem to occur owing to disrupted embryogenesis beginning as early as the fourth week of embryonic life.[1] The diagnosis is established using cardiac imaging studies, such as transthoracic echocardiography or left ventricular angiography, after exclusion of coronary artery disease, cardiomyopathies, and other inflammatory or traumatic causes.[2-7] Most patients with congenital left ventricular aneurysms and diverticula are clinically asymptomatic. However, patients may present with cardiac arrhythmias, embolic complications, or heart failure.[8]

There are only few observational case reports and small series on congenital left ventricular aneurysms and diverticula in the medical literature.[9-14] The association of this abnormality with ventricular tachyarrhythmia was reported in 1971 by Maloy et al.[9] However, very little is known on the clinical outcome of these patients to date. In this study, we aimed to investigate the long-term outcome of patients with congenital left ventricular aneurysms and diverticula who had ventricular arrhythmias or presyncope/syncope at initial presentation.

CLINICAL SIGNIFICANCE

- Ventricular tachycardia in patients with congenital left ventricular aneurysm or diverticula is often monomorphic and has a right bundle branch block morphology.
- In patients with congenital left ventricular aneurysm or diverticula, the clinical ventricular tachycardia is usually inducible during electrophysiologic study, indicating a role for this test in risk stratification.
- In some patients the implantation of an implantable cardioverter defibrillator is required to prevent life-threatening ventricular tachyarrhythmias.

METHODS

We retrospectively screened hospital charts dated 1990 and after for patients diagnosed with congenital left ventricular aneurysms and diverticula in the Clinic for Cardiology, University Hospital, Zürich, Switzerland. The diagnosis of congenital left ventricular aneurysms and diverticula was made using transthoracic echocardiography after exclusion of coronary artery disease, local cardiac inflammatory process, or traumatic causes, as well as cardiomyopathies in all patients. Patients with diverticula or aneurysms in cardiac chambers other than the left ventricle were also excluded from the analysis.

All patient records were screened for demographic and clinical data, 12-lead echocardiogram (ECG) findings, transthoracic ECGs, 24-hour Holter recordings, and electrophysiologic studies, if available. Data available from hospital charts and those obtained from referring physicians on the clinical management of these patients, including pharmacologic treatment, catheter ablation, and implantation of an implantable cardioverter defibrillator, were collected. The clinical outcome of these patients was assessed until the date of last follow-up.

On transthoracic echocardiography, a left ventricular *aneurysm* was diagnosed in the presence of a protrusion from the left ventricular cavity, typically with a wide base, not necessarily contracting in synchrony to the left ventricle (depending on the amount of myocardial involvement). In contrast, a left ventricular *diverticulum* was characterized by a finger-like protrusion with a narrow base, which contracts in synchrony with the ventricular chamber.

All patients in the study cohort presented clinically with arrhythmic manifestations at baseline. This included ECG documentation of ventricular arrhythmias or ectopic beats, or clinical symptoms suggesting the presence of an underlying arrhythmia, such as palpitations or presyncope and/or syncope.

RESULTS

A total of 250 patients had the diagnosis of congenital left ventricular aneurysms and diverticula in our institution. In this cohort of patients, 30 had ventricular arrhythmias or presyncope and/or syncope at initial presentation. Characteristics of these patients are provided in **Table 1**. The study cohort had a mean (±SD) age of 46.6 ± 19.4 years. Twenty-one patients (70%) were male. Two patients had a family history of sudden death in the absence of a known cardiac disorder (patients 8 and 20).

On transthoracic ECG, the mean left ventricular ejection fraction was 57% ± 8.7%. The congenital left ventricular aneurysms and diverticula had a posterobasal localization in 12, apical in 9, anteroseptal in 4, and anterolateral in 5 patients (**Figures 1** and **2**). Six patients presented with more than 1 left ventricular aneurysm and diverticulum.

Arrhythmic Manifestations at Presentation

One patient had survived sudden cardiac death with documented ventricular fibrillation at initial presentation (patient 5). Seven patients (23%) presented with a ventricular tachycardia at baseline. Six patients (20%) had symptomatic ventricular ectopic beats. The remaining patients presented with presyncope and/or syncope.

Arrhythmias During Follow-Up

Multiple 12-lead ECGs and 24-hour Holter recordings revealed spontaneous ventricular tachycardia in 17 patients (57%) with congenital left ventricular aneurysms and diverticula; 13 of these had a sustained ventricular tachycardia. The ventricular tachycardia was monomorphic in 9 patients (**Figure 3**), polymorphic in 4 patients, and both monomorphic and polymorphic in 1 patient. Eight patients